# HCMJ

## HILL COUNTRY MEDICAL JOURNAL 2023

*"Optimal patient care amidst challenging times"*

Volume 5 - Issue 1 - January 2023
ISSN 2613 - 8581

# 12th Annual Academic Sessions

## 18th and 19th January 2023

HCCS ACADEMIC SESSIONS | REVIEW ARTICLE | RESEARCH ARTICLES | CASE REPORTS & SERIES

Published by Hill Country Clinical Society

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# 12th Annual Academic Sessions
# 18th and 19th January 2023

## Programme Book

**Edited by**
**Dr. Thaha M Musthafa**
**Dr. Lakshman Bandara**

**Published by Hill Country Clinical Society**

# Contents

# Message from the President

I am very much honored and privileged to write here as the 12th President and youngest president of the Hill Country Clinical Society since its establishment in 2010. The HCSS is the pioneer of clinical excellence in Nuwara Eliya district where it was able to uplift the knowledge, practice and skills of Medical professionals in this under privileged district of Sri Lanka.

This 2022/2023 is a challenging to everyone in Sri Lanka with a series of crisis –COVID 19 pandemic, political instability - started few years ago and still continuing with the ongoing economic crisis while slowly returning to a new normal life. With the theme of "Optimal Patient Care Amidst Challenging Times" we as the Council of 2022/23 organizing the most prestigious event of the year, HCCS Annual Academic Sessions as a two days event.

This year we were able to conduct numerous academic activities despite crisis situation. Uninterrupted CME lectures with CME Points awarded from Sri Lanka Medical Association accreditation every fort night. Further we were able to organize many events in Nuwara Eliya with the all four mail colleges and other national and local Professional organizations. Sri Lanka College of Obstetricians and Gynecologists first Regional Workshop (3 days), Sri Lanka College of Paediatricians first Regional workshop (2 days) and also with other colleges such as Sri Lanka Society of Breast Surgeons, Anti Leprosy campaign, and many others.

Moving to paper free, ecofriendly movement, we introduced e- certificate to participants of the academic activities. This is I consider as a huge achievement in HCCS history and will be continue in future as well.

Not only academic activities, this year we were able to organize non-academic activities as the first such event in the history. Also, this year we have planned 2 pre congress workshops in collaboration with prestigious Peradeniya Medical School Alumni Association (PeMSAA) to doctors and nurses for the first time expanding our expertise to other health care sector development. This event will enlighten the history of HCCS were the annual academic sessions have expanded to 2 days event.

In parallel to annual academic sessions, we publish our Hill Country Medical Journal (Volume 5, Issue 01) which we are going to make a bi-annual journal soon which will encourage doctors in the region to encourage conducting research and providing a platform to publish their scholastic activities.

Annual Academic Sessions is the main academic event in our calendar year which will be on 18th and 19th January 2023. I take this opportunity to welcome our Chief Guest Dr H T Wickramasinghe, Guest of Honor Prof Thushara Kudagammana, invited guests and resource personals who are coming all the way to Nuwara Eliya from all over the country. Also I would like to invite all the medical professionals to join our climax event and to embrace the natural beauty of Nuwara Eliya.

Finally, this year events will not be a success without my Council of Hill Country Clinical society, Colleague Consultants, junior medical officers and office staff. I thank all of them for guiding, supporting and being with me this challenging time and for their tireless work for the successful end of the 2022/2023 tenure.
I hope HCCS will flourish with blessings of all the past presidents, past councils and will the upcoming president and council will take further high of the society. I wish all the success of annual sessions and future activities of HCCS.

**Dr Jagath Ranasinghe MBBS, DCH, MD (Paediatrics), FRSPH (UK)**
Consultant Paediatrician
President 2022/23
Hill Country Clinical Society

## Message from the Director, DGH - Nuwara Eliya

It is with great pleasure, I am writing this message as the Director of the Hospital and the Patron of the HCCS to 12th annual academic session's year 2022.

As the apex hospital for the Nuwara Eliya district, DGH Nuwara Eliya provides health care services to a population of more than 800000 people, mainly from Nuwara Eliya district, as well as from adjacent Badulla and Kandy districts. In addition to local residents, the institute also serves the visitors from all over the country and foreign countries who visit Nuwara Eliya.

The addition of a 650-bedded modern new hospital building complex with up-to-date equipment and facilities is a major achievement in the hospital history.

The introduction of the concept of Clinical Governance is a very important step taken to get the best use of these facilities and to achieve excellence in clinical care. As I have experienced in the United Kingdom, CG plays a major role in achieving clinical excellence in NHS UK and other developed countries.

Even though there is no designated CG programme in our country, some activities and practices conducted by the HCCS of DGH Nuwara Eliya are comparable to the CG programme in United Kingdom. Continuous Professional Development, clinical audits, staff welfare, helping hospital research and audit committee, hospital ethical committee, patient and public involvement, incident reporting and risk management and implementation of HHIMS system are the main concepts we have already adopted in the introduction of CG in DGH Nuwara Eliya. With that, we have achieved a significant improvement in the quality, safety and productivity of the patient care.

I highly appreciate HCCS on its involvement in better patient care and I would like to congratulate the President and the council members at their success in organizing academic and non-academic activities.

I wish success to HCCS for their future academic and non-academic activities.

**Dr Mahendra Senevirathna**
Director
District General Hospital
Nuwara Eliya

## Message from the Chief Guest

It is a great privilege and a pleasure to me to be the chief guest of the 12th Annual academic sessions of The Hill Country Clinical Society (HCCS). The theme of this year "Optimal patient care amidst challenging times" is a very timely and appropriate theme in the context of present crisis in our country.

The work done by The HCCS over last 12 years in the field of medicine and health care is praiseworthy. The modern philosophy of evidence based medicine is to guide the medical professionals to give the best care possible to patients using the best available research evidence. Educating own colleagues is an integral component of this endeavor.

The HCCS is performing this duty well within their boundaries and available resources. In fact they have gone beyond encompassing public education as part of their responsibility. I have seen them using social media very successfully to reach the public, which is not yet seen as a regular feature with other clinical societies in the country. I wish The HCCS will have the strength and courage to continue its efforts to promote healthcare and health education in the region. I wish all the success in this 12th Annual session of your society.

**Dr. H T Wickramasinghe**
Consultant Paediatrician
Neville Fernando Teaching Hospital
Sri Lanka

## Message from the Guest of Honour

It is my pleasure and honor to be the invited as the Guest of Honor at the 12th annual sessions of the Hill Country Clinical Society, which is the premier clinical society in the hill country geared to disseminate new knowledge to health care professionals in the region.

The field of medicine along with other sciences is developing at a breathtaking pace. Keeping up with the developments is a difficult but an essential component of patient care services specially under the current array of crises. The patient demands are ever increasing for a better health care with easy access to knowledge at their fingertips. Unless the clinicians keep with this pace, the patients get an unfair deal; the clinicians too at the risk of being left behind. The Hill country clinical society is performing a herculean task by getting the health care providers engaged, motivating them and providing them with necessary knowledge and skills for better patient care delivery.

We live in an era of evidence based medicine. While delivering patient care services with up to date knowledge, it is also important to contribute to evidence based medicine as well. This is achieved by engaging in research. Though 4/5th of world population is in the developing world, it is the developed world which contributes to 4/5th of the world research. I invite the members of this august society to contribute to research, utilizing the wealth of clinical material at your disposal. Getting in touch with university academics in the region will certainly help to overcome the barriers to engage in research.

I would like to congratulate the president and the council for dishing out an excellent academic programme and wish all the very best for a fruitful academic session.

I would like to congratulate the president and the council for dishing out an excellent academic program and wish all the very best for a fruitful academic session.

**Prof. Sanath T. Kudagammana**
Professor in Paediatrics
Faculty of Medicine
University of Peradeniya

## Message from the Joint Secretaries

We were distinguished to be the joint secretaries of the Hill Country Clinical Society, which is the foremost academic body of Nuwara Eliya District. Moving on with the role of joint secretaries we have coordinated clinical education programmes under the guidance of the honorable president whose utmost dedication always led to the path of success of our activities.

Through our past year we have commenced many academic activities and non-academic activities, including CME sessions which held every fortnight. We hope our academic programmes nurtured knowledge and guidance for health care professionals to improve their skills. However, as every beginning has an ending, at the end of a successful & eventful year, we are at the apex event of the Hill Country Clinical Society; that is the 12$^{th}$ Annual Academic Sessions.

Hill Country Medical Journal which is published annually is the masterpiece of important information gathered via original researches, case reports, reviews and other scholastic activities from Nuwara Eliya as well as from other regions.

The editors have been working tirelessly in bringing up the standard of HCMJ.

Finally we would like to thank our honorable president Dr. Jagath Ranasighe for his enormous work throughout the year despite the challenges we faced over the past year including Sri Lanka's huge economic crisis. We are also grateful for all the council members, consultants and medical officers who gave their support in making the events successful.

**Dr. Malitha Hewage and Dr. Kalumini Weheragoda**
Joint Secretaries
Hill Country Clinical Society

## Message from the Editors

The Hill Country Clinical Society is the apex academic body in the Nuwara Eliya district. We have been organizing numerous CME activities with esteemed resource persons from many parts of the island. As a summit of the academic events, we are treading past our 12th annual academic sessions with the theme "Optimal Patient Care Amidst Challenging Times".

Breaking through the Covid-19 barrier and socioeconomic crisis we tried to promote the true meaning of our theme by uplifting the patient care while exhibiting love, concern, sensitivity, kindness and tolerance at all the time. The annual academic event witnesses Workshops, Plenaries, Symposiums and hands on Skills sessions for doctors and nurses, without any doubts will upgrade the knowledge and the skills of the health care providers.

This journal comprises scientific writing from many facets of medicine and we strongly believe that these will ensure exchange of academic knowledge towards "Evidence Based Medicine". We appreciate the hard work and dedication by all the authors and our organizing committee members for their tireless work towards the success.

We wish all of you a remarkable academic experience and an enjoyable conference during 12th Annual Academic Sessions of HCCS .

**Dr. Thaha M Musthafa**
**Dr. Lakshman Bandara**
Co - Editors
Hill Country Clinical Society

# HILL COUNTRY CLINICAL SOCIETY COUNCIL 2022/2023

| | |
|---|---|
| ***President*** | Dr Jagath Ranasinghe |
| ***Vice President*** | Dr Sonali Gunathilaka |
| ***Immediate Past President*** | Dr Sandeepana Gamage |
| ***Patron*** | Dr Mahendra Senevirathna |
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| ***Assistant Treasurer*** | Dr Subuddhika Illukkumbura |
| ***Co Editors*** | Dr Thaha M Musthafa |
| | Dr Lakshman Bandara |
| ***Editorial Board*** | Dr Bingumal Jayasundara |
| | Dr Ravi Dissanayaka |
| | Dr Sonali Gunathilaka |
| | Dr. Jagath Ranasinghe |

***Committee Members***

| | |
|---|---|
| Dr Saman Rajaguru | Dr Thushara Ranasundara |
| Dr Kamal Rajaguru | Dr Deepal Nanayakkara |
| Dr Duleeka Dissanayaka | Dr Rukshan Dissanayake |
| Dr Ranga Gunasekara | Dr Gayan Rajapaksha |
| Dr Paboda Samarathunga | Dr Kavisha Dissanayaka |
| Dr Kusala Sandamalee | Dr Prasad Abeyrathne |
| Dr Chandani Mendis | Dr Chamara Dematawa |
| Dr Chamindi Seneviratne | Dr Manjula Herath |
| Dr Shyama Yapa Bandara | Dr Sajini Athurupana |
| Dr Anitha Rajendra | Dr Prasanna Wijerathne |
| Dr Rasangika Gunasekara | |

# HILL COUNTRY CLINICAL SOCIETY
## DISTRICT GENERAL HOSPITAL- NUWARA ELIYA
## COUNCIL AND ORGANISING COMMITTEE 2022/2023

**Seated Left to Right**

Dr. Kalumini Weheragoda (Joint Secretary), Dr. Anuruddha Wijesooriya (Treasurer), Dr. Lakshman Bandara (Co Editor), Dr. Jagath Ranasinghe (President), Dr. Mahendra Seneviratne (Patron), Dr. Sonali Gunatilake (Vice President) Dr. Ranga Gunasekara, Dr. Thaha Musthafa (Co Editor), Dr. Subuddhika Illukkumbura (Assistant Treasurer), Dr. Malitha Hewage (Joint Secretary)

**Standing 1st Raw Left to Right**

Dr. Dinuka Munasinghe, Dr. Anusha Gardiawasam, Dr. Rasangika Gunasekara, Dr. Sajini Athurupana, Dr. Thushara Ranasundara, Dr. Chamindi Senevirathne, Dr. Paboda Samarathunga, Dr. Anitha Rajendra, Dr. Kavisha Dissanayaka, Dr. Kusala Sandamalee, Dr. Chandanie Mendis, Dr. Shyama Yapa Bandara, Dr. Gayan Rajapaksha

**Standing 2nd Raw Left to Right**

Dr. Hemantha Dissanayaka, Dr. Ravi Dissanayake, Dr. Prasad Abeyrathne, Dr. Chamara Dematawa, Dr. Deepal Nanayakkara, Dr. Lasith Obodarachchi, Dr. Kameera Bopaththa, Dr. Saman Rajaguru, Dr. Manjula Herath, Dr. Rukshan Dissanayaka, Dr. Kamal Rajaguru, Dr. Duleeka Dissanayake

# ACADEMIC AND NON ACADEMIC ACTIVITIES OF HCCS 2022

| DATE | EVENT | RESOURCE PERSON |
|---|---|---|
| **02/02/2022** | **DIABETES UPDATE**<br>***(CME LECTURE)*** | **Dr. Sonali Gunatilake**<br>**Consultant Endocriniologist**<br>**DGH – Nuwara Eliya** |
| **24/02/2022**<br>**25/02/2022**<br>**26/02/2022** | **SAFE MOTHERHOOD WORKSHOP**<br>***In Collaboration with Sri Lanka College of Obstetricians and Gynaecologists*** | **Day One - Inauguration**<br>**Day Two - Hands on skills**<br>**Day Three- Lectures** |
| **14/03/2022** | **INAUGURATION OF BREAST CARE CLINIC & TRAINING ON BREAST EXAMINATION**<br>***In collaboration with National Cancer Control Programme (NCCP) and Sri Lanka Society of Breast Surgeons (SLSBS)*** | **Faculty from NCCP and SLSBS and DGH Nuwara Eliya** |
| **16/03/2022** | **SUDDEN ONSET SENSORY NEURAL HEARING LOSS - AN ONTOLOGICAL EMERGENCY**<br>***(CME LECTURE)*** | **Dr. Jayantha Bandara Kallora**<br>**Consultant ENT Surgeon**<br>**DGH - Nuwara Eliya** |
| **30/03/2022** | **COMMEMORATING WORLD LEPROSY DAY 2022 – AWARENESS PROGRAM**<br>***In collaboration with Anti-Leprosy Campaign , Ministry of Health and RDHS Nuwara Eliya*** | **Dr. Chathurarya Siriwardena**<br>**Consultant Dermatologist**<br>**DGH – Nuwara Eliya** |
| **27/04/2022** | **HOW DO YOU PROTECT THE PATIENT ONCE HEAD INJURY HAPPENED?**<br>***(CME LECTURE)*** | **Dr. Shermila Gayathri**<br>**Consultant Anaesthetist**<br>**DGH – Nuwara Eliya** |
| **11/05/2022** | **HYPOKALAEMIA, WHAT DO YOU NEED TO KNOW?**<br>***(CME LECTURE)*** | **Dr. Paboda Smarathunga**<br>**Consultant Physician**<br>**DGH – Nuwara Eliya** |
| **25/05/2022** | **IMAGING BASICS IN CLINICAL PRACTICE**<br>***(CME LECTURE)*** | **Dr. Kusala Sandamali**<br>**Consultant Radiologist**<br>**DGH – Nuwara Eliya** |
| **10/06/2022** | **WORKSHOP ON CLUBFOOT AND THE PONSETI METHOD** | **Dr.Malinda Ileperuma**<br>**Consultant Orthopaedic Surgeon**<br>**DGH – Nuwara Eliya** |
| **24/06/2022** | **AN UPDATE ON SEPSIS**<br>***(CME LECTURE)*** | **Dr. Kavisha Dissanayake**<br>**Consultant Anaesthetist**<br>**DGH – Nuwara Eliya** |
| **07/07/2022** | **DIFFERENTIATING HIP PATHOLOGY FROM LUMBAR SPINE PATHOLOGY;A DIAGNOSTIC CONUNDRUM** | **Dr.Malinda Ileperuma**<br>**Consultant Orthopaedic Surgeon**<br>**DGH – Nuwara Eliya** |
| **14/07/2022** | **MANAGEMENT OF FEMALE BREAST CANCER *(CME LECTURE)* – LESSONS LEARNT FROM EXPERIENCES AT NUWARA ELIYA** | **Dr. Bingumal Jayasundara**<br>**Consultant General Surgeon**<br>**DGH – Nuwara Eliya** |
| **28/07/2022** | **THEY ARE AMONG US; AN INSIGHT INTO TRANSGENDER CARE**<br>***(CME LECTURE)*** | **Dr. Sonali Gunatilake**<br>**Consultant Endocriniologist**<br>**DGH – Nuwara Eliya** |

| | | |
|---|---|---|
| **02/08/2022** | **EMBRACE THE NATURE WITH AN ENVIRONMENTALIST** | **Dr. Thilak Kandegama<br>Environmentalist<br>Seehela Native Farming Education Centre** |
| **03/08/2022** | **SEXUAL DYSFUNCTIONS IN MALES**<br>***(CME LECTURE)*** | **Dr. Thaha M. Musthafa<br>Consultant Genito Urinary Surgeon<br>DGH – Nuwara Eliya** |
| **22/08/2022** | **INAUGURATION OF "MULTIDISCIPLINARY CLEFT PALATE CENTER" at DGH Nuwara Eliya**<br>***In collaboration with Multidisciplinary Cleft Palate Center Teaching Hospital Karapitiya*** | **Faculty From Teaching Hospital Karapitiya<br>Faculty of Dental Sciences Peradeniya<br>OMF & Orthodontic Unit DGH – Nuwara Eliya** |
| **31/08/2022<br>01/09/2022** | **SLCP REGIONAL SESSIONS**<br>***In collaboration with Sri Lanka College of Paediatricians*** | **Faculty from SLCP & DGH – Nuwara Eliya<br>Day One - Inauguration<br>Day Two - Lectures** |
| **08/09/2022** | **LIVE WEBINAR TELECAST OF UPDATE ON MANAGEMENT OF COMMON NCDs**<br>***Conducted by Sri Lanka College of Internal Medicine in collaboration with Ministry of Health*** | **Faculty from College of Internal Medicine** |
| **12/10/2022** | **DERMATOLOGY QUIZ OF THE YEAR**<br>***(CME LECTURE)*** | **Dr. Tharindu Dinupa<br>Consultant Dermatologist<br>DGH – Nuwara Eliya** |
| **26/10/2022** | **CEILING OF CARE**<br>***(CME LECTURE)*** | **Dr. Ranga Gunasekara<br>Consultant Physician<br>DGH – Nuwara Eliya** |
| **09/11/2022** | **WORLD HEART DAY COMMOMORATION PROGRAMME - AWARENESS ON HEART ATTACKS AND NON COMMUNICABLE DISEASES** | **Dr. Lakshman Bandara<br>Consultant Cardiologist<br>DGH – Nuwara Eliya** |

# Pre Congress Workshops

## Pre Congress Workshop for Doctors {IN COLLABORATION WITH Peradeniya Medical School Alumni Association (PeMSSA)}
## "Approach to Common Clinical Emergencies"

18TH JANUARY 2023
**09:00 – 13:00**
At New Auditorium, District General Hospital – Nuwara Eliya

| | | |
|---|---|---|
| **Emergency Quiz** | Dr. Kameera Bopaththa | Consultant Emergency Physician, District General Hospital - Nuwara Eliya |
| **Neonatal Emergencies** | Dr. Thushara Ranasundara | Consultant Neonatologist, District General Hospital- Nuwara Eliya |
| **Approach to Shock** | Dr. Kameera Bopaththa | Consultant Emergency Physician, District General Hospital - Nuwara Eliya |
| **Postpartum Haemorrhage** | Dr. M Champika Gihan | Senior Lecturer - Faculty of Medicine, University of Peradeniya<br>Consultant Obstetrician & Gynaecologist, |
| **Upper GI Bleeding** | Dr. Buddhika Dassanayake | Consultant Surgeon, Senior Lecturer – Department of Surgery, Faculty of Medicine University of Peradeniya |
| **Paediatric Emergencies** | Dr. Shyama Yapa Bandara | Consultant Paediatrician, District General Hospital - Nuwara Eliya |
| **Hands on Skills** | Interactive session | All Faculty |

## PRE-CONGRESS WORKSHOPS FOR NURSES

18TH JANUARY 2023
**09:00 – 12:30**
At Old Auditorium, District General Hospital – Nuwara Eliya

### "APPROACH TO A DETERIORATING PATIENT"

Opening remarks
**"A Deteriorating Patient"**
**"Transporting a Critically Ill Patient"**
**"Basic Airway Management and Oxygen Therapy"**
**"Fluids and Tubes"**

Course Directors
Dr. Ranga Gunasekara – Consultant Physician, District General Hospital - Nuwara Eliya
Dr. Kavisha Dissanayake - Consultant Anaesthetist, District General Hospital - Nuwara Eliya

# HCCS – 12TH ANNUAL ACADEMIC SESSIONS
# MAIN CONGRESS PROGRAMME

19TH JANUARY 2023

At New Auditorium, District General Hospital – Nuwara Eliya

| | | |
|---|---|---|
| **08:30 - 08:45** | Registration | |
| **08:45 - 09:00** | Procession of Guests<br>National Anthem<br>Lighting of Oil Lamp | |
| **09:00 – 09:05** | Welcome Address by President - Hill Country Clinical Society – **Dr. Jagath Ranasinghe** | |
| **09:05 – 09:10** | Address by Director - District General Hospital - Nuwara Eliya - **Dr. Mahendra Senevirathne** | |
| **09:10 – 09:35** | Address by the Chief Guest - ***Is Evidence Based Medicine, The Gold Standard in Clinical Practice or a Hype?"*** - **Dr. H T Wickramasinghe -** Consultant Paediatrician, Neville Fernando Teaching Hospital – Colombo | |
| **09:35 – 10:00** | Address by the Guest of Honour – ***"Chronobiology and its implication in disease"*** - **Prof. S T Kudagammana -** Professor in Paediatrics - Faculty of Medicine University of Peradeniya, President- Peradeniya Medical School Alumni Association (PeMSAA) | |
| **10:00 - 10:20** | **Morning Tea** | |
| **10:20 - 10:40** | **Plenary Number 01 - "Morbid Jealousy- An Under-Recognized Cause of Domestic Violence" Dr. L L Amila Isuru** Senior Lecturer, Faculty of Medicine, Rajarata University and Honorary Consultant Psychiatrist, Teaching Hospital - Anuradhapura | |
| **10:40 - 11:30** | **Symposium Number 01 - "Different Dimensions of Pulmonary Hypertension- Case Based Discussion"** | |
| | Panelist 1 | **Dr. Ranga Gunasekara -** Consultant Physician, District General Hospital - Nuwara Eliya |
| | Panelist 2 | **Dr. Lakshman Bandara -** Consultant Cardiologist, District General Hospital - Nuwara Eliya |
| | Panelist 3 | **Dr. Prasanna Wijerathne -** Consultant Respiratory Physician, District General Hospital - Nuwara Eliya |
| **11:30 - 11:55** | **Plenary Number 02 - "Beauty of Breast Milk" - Dr. H T Wickramasinghe** Consultant Paediatrician, Neville Fernando Teaching Hospital – Colombo | |
| **11:55 - 12:45** | **Symposium Number 02 - "Wound care: Bench to Bedside"** | |
| | 1. Introduction and Basics | **Dr. Prasad Abeyratne -** Consultant Surgeon, District General Hospital - Nuwara Eliya |
| | 2. Principles of Wound healing and dressing, Pressure Ulcers | **Dr. Pragaash Shanmuganathan** - Consultant Plastic and Reconstructive Surgeon, Sirimavo Bandaranayake Specialized Children's Hospital - Peradeniya |
| | 3. How to Approach Hard-to-Heal wounds | **Dr. B A Donald Rubakan** - Consultant Vascular and Transplant Surgeon, Teaching Hospital - Anuradhapura |
| **12:45 - 13:25** | **Lunch** | |
| **13:25 - 13:45** | **Plenary Number 03 – "Renal Replacement Therapy, When? How? And To Whom?** **Dr. Premil Rajakrishna –** Consultant Nephrologist, Teaching Hospital - Kurunegala | |
| **13:45 - 14:35** | **Symposium Number 03 - "Facial Aesthetic – Where art meets science"** | |
| | 1. Facing the Face: Art and science of Facial Sculpting | **Dr. Buddhika Dassanayake** - Consultant OMF Surgeon, Provincial General Hospital – Badulla |
| | 2. Braces beyond the teeth – Role of Orthodontics in Facial Aesthetics | **Dr. Deepal Nanayakkara -** Consultant Orthodontist, District General Hospital - Nuwara Eliya |
| | 3. New Generation of Medical Aesthetics | **Dr. Tharindu Dinupa -** Consultant Dermatologist and Lecturer Faculty of Medicine, University of Moratuwa |
| **14:35 - 14:50** | Quiz Session with Prizes | |
| **14:50 - 15:00** | Vote of Thanks - Secretary - Hill Country Clinical Society | |

## ACADEMIC PROGRAMME - FACU LTY

**Dr. L L Amila Isuru**
Senior Lecturer,
Faculty of Medicine, Rajarata University.
Honorary Consultant Psychiatrist,
Teaching Hospital,
Anuradhapura

**Dr. Pragash Shanmuganathan**
Consultant Plastic and Reconstructive Surgeon
SBSCH,
Peradeniya

**Dr Ranga Gunasekera**
Consultant Physician,
District General Hospital,
Nuwara Eliya

**Dr. B A Donald Rubakan**
Consultant Vascular and Transplant Surgeon,
Teaching Hospital,
Anuradhapura

**Dr Lakshman Bandara**
Consultant Cardiologist,
District General Hospital,
Nuwara Eliya

**Dr. Premil Nadeekanth Rajakrishna**
Consultant Nephrologist,
Teaching Hospital,
Kurunegala

**Dr. Prasanna Wijerathne**
Consultant Respiratory Physician,
District General Hospital,
Nuwara Eliya

**Dr. Deepal Nanayakkara**
Consultant Orthodontist,
District General Hospital,
Nuwara Eliya

**Dr. H T Wickramasinghe**
Consultant Paediatrician,
Neville Fernando Teaching Hospital,
Colombo

**Dr. Buddhikka Dassanayake**
Consultant Oral Maxillofacial Surgeon,
Provincial General Hospital,
Badulla

**Dr. Prasad Abeyratne**
Consultant Surgeon,
District General Hospital,
Nuwara Eliya

**Dr. Tharindu Dinupa**
Lecturer / Consultant Dermatologist,
Faculty of Medicine,
University of Moratuwa

# HCMJ

## HILL COUNTRY MEDICAL JOURNAL 2023

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The Editor, Hill Country Medical Journal
Hill country clinical society office
District General Hospital
Nuwara Eliya
Email;
hillcountryclinicalsocietyeduc@gmail.com



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**Cover Photo**
Rhino-horned Lizard
*(Ceratophora Stoddartii)*

Photo Credit
Dr Chathura S. Gunadasa

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## Editorial



## Article Review



## Research Articles



## Case Reports

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*Dharshini Karuppiah*
21. Non-Traumatic Depressed Skull Fracture with Epidural Haemorrhage in a Neonate
*Dharmawardhana P M , Ekanayake E M G K, Hewage M N, Ranasundara T M, Nathawitharanalage K S*
22. Story behind a Granulomatous Facial Lesion - Angiosarcoma the Hidden Monster
*Dinupa KDT, Fernando WKBA, Perera WNC Abeygunasekara Priyanka*
23. Siblings Diagnosed With Papillon-Lafevre Syndrome; One Presented With Recurrent Liver Abscess; The Other Presented With Hyperkeratosis and Skin Infection
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24. Incidentally Found Mesenteric Pseudocyst of the Ileum
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# "OPTIMAL PATIENT CARE AMIDST CHALLENGING TIMES"

**Dr. Thaha M Musthafa, Dr. Lakshman Bandara**

Year 2022 was the year of moving out of the pandemic when the whole world starts breathing freely by defeating the SARS-Cov2; COVID-19 infection. During the pandemic, aside from the devastating loss of lives, people around the country had faced an unprecedented loss of incomes, livelihoods and elevated threats of poverty. However, Sri Lankan health care sector played a major role in the control of COVID-19 infection by practicing strict adherence to infection control policies and strengthening the vaccination programme throughout this difficult period. Although the whole healthcare system was under immense stress, finally Sri Lankan health care team was able to lead the country to move out from the pandemic despite of several sacrifices.

Once COVID-19 pandemic hits the country, the Sri Lankan economy was at a particular low ebb, having persistent low economic growth and a high and escalating public debt burden. Therefore, the pandemic had a strong impact on destabilization of the Sri Lankan economy in last few years and the country's economic growth outlook in 2022 had been marked by bouts of unsteadiness, which was mainly linked to public unrest and political instability of the country.

The unsolidified political situation and heightened monetary, external and financial sector imbalances had posed significant uncertainty for Sri Lanka's economic outlook. Key downside risks which were included a slow debt restructuring process, limited external financing support, and major negative hits on tourism, which leads to prolong recovery from the scarring effects of the health, economic and political crisis. This had made a significantly negative impact on the country's health sector as well.

Primarily, the economic crisis has made the national health care system in jeopardy. Health-care workers were attempting to keep the Sri Lankan health-care system stable in various aspects. However, fuel shortage, interrupted supply of medical equipment and shortages of essential medicine had made year 2022, critical for both patients and health-care workers.

Many of the observation in rural community prove that the people had given priority to food and shelter than health in the background of continued higher inflation of the country. Addition to that, the access to available health care system was also affected in numerous aspects due to higher transport cost and rising poverty. Without a doubt, the economic crisis majorly affected the mental health of Sri Lankan people as well. Schools have closed frequently and regular power disruptions have made online learning difficult. Though these elements were less addressed, the crisis had a major impact on the mental health of the society. We experience the gravity of the problem nowadays as a tip of the iceberg.

In general, year 2022 was a challenging year to every citizen and the health care system was severely affected in each

strata. However, many of new strategies were came up to overcome these challengers. We, as Hill Country Clinical Society (HCCS) took some initiatives to obtain foreign funds to purchase essential medicine and medical equipment to our rural local community. Many of the foreign donors were coordinated via the HCCS as a trusted channel. More importantly, though health care team faces many obstacles, the routine duty on behalf of the patients was minimally affected due to their utmost dedication. Addition to that the system is still able to maintain the infectious disease prevention, continue the emergency services and routine care for non-communicable diseases while maintaining the primary health care system with minimal interruptions.

In year 2023 should be a new era to overcome the health and economic challenges and to deliver optimal medical care admits this crisis situation.

It is highlighted that it is essential to re-organize the primary health care system to meet Sri Lanka's future needs; using the experience during the crisis situation to improve people-centered service provision. Especial emphasis should be made to eliminate the wastage in health care system and to implement appropriate human resource management.

Additional attention should be focused on improving the efficiency and reliability of supply chain management systems for pharmaceuticals and essential medical supplies. The adverse experience that we had during the crisis situation should be a good lesson for the future.

It is also highlighted that the policy makers should continually invest and expand the country's human resources despite of "brain drain'' in the health care system during these crisis situation. If this factor is not addressed apriority, availability of skillful human resources which is necessary to provide quality, people-centered health care to Sri Lankans will be affected in near future.

As we were moving out of the COVID-19 pandemic in 2021 and, stepping through a political and economic crisis in 2022 with rising challenges day by day, still Sri Lanka has the potential to provide healthcare for all its citizens in a way that is effective, affordable, and accessible with the dedication of all health care team.

# REMISSION OF DIABETES: MYTH OR A REALITY

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**Abstract**

Type 2 diabetes mellitus (T2D) was known as a progressive chronic disease with an alarming prevalence worldwide. complications. Despite improvement of treatment options and care in diabetes, diabetes remains a huge burden to the healthcare system. Remission has recently emerged as a therapeutic target in T2D, achievable through a wide range of interventions. Recent studies have shown that extensive lifestyle changes, such as dietary restrictions, weight reduction, bariatric surgery, and intensive glucose lowering therapy, can prompt the remission of diabetes, but some unanswered questions remain regarding its long-term remission rates and effects on diabetic complications. In this review, available therapeutic approaches to target the remission of diabetes is discussed.

**Key words** – Remission, Reversal, Diabetes, Low calory, Low carbohydrate, Bariatric surgery, Weight loss, Diet

## Introduction

Diabetes has become an epidemic in the society with a prevalence of 537 million adults living with diabetes according to the International Diabetes Federation data (1). Of them, 90 million adults are living with diabetes in South Asia which is projected to be 152 million by 2045 consuming a significant amount of health budget to facilitate treatment (1).

Type 2 diabetes mellitus (T2D) is considered as a chronic progressive disease with a significant mortality and morbidity due to associated macro- and microvascular complications as well as hyperglycemia. Treatment strategies are aimed at managing glycemic levels and reducing the occurrence of complications.

It is evident that pre-diabetes is reversible through targeted lifestyle modifications and/or medications like Metformin but until recent, reversal of diabetes once diagnosed was not a discussion on the table (2-4). With recent evidence, chronic irreversible nature of T2D has been challenged by strategies targeting "Reversal" of diabetes to achieve normoglycemia. (5).

In 2009, American Diabetes Association (ADA) in its consensus statement had discussed about diabetes remission and in 2016, the World Health Organization (WHO) indicated the possibility of reversal of diabetes quoting Barbados diabetes reversal study (6, 7). Above together with landmark clinical trials, interventional studies and real-world experience opened eyes of the world in to the possibility of remission or reversal of type 2 diabetes (8, 9). Issue of recent consensus report on diabetes remission by ADA has also shed some light in to the place and importance of diabetes remission although standard guidelines on T2D still have not incorporated strategies targeting diabetes remission as standard of care (10). However, with current evidence, it is evident that T2D is metabolically reversible (8, 9, 11).

## Terminology

Terms remission, reversal and cure had been used in the literature at different

levels with regard to T2D. However, *Remission* of T2D is considered more appropriate as it denotes no active evidence of hyperglycemia, yet the improvement of glycemic state would not be permanent. *Reversal* of T2D indicates process of returning to glycemic levels below those diagnostic of diabetes, yet it should not be equated with the state of remission (10). *Cure* is a state where the pathology has normalized and no further follow-up or management is needed. As there is the potential for re-occurrence of T2D which is documented in the literature and where current evidence is not sufficient to conclude cure of T2D, use of the term cure may give a false re-assurance (12).

**Definition of Remission of T2D**

ADA defined remission of diabetes as glycated hemoglobin (HbA1c) < 6.5% (48 mmol/mol) or fasting blood glucose (FPG) < 126 mg/dL (7 mmol/L), or estimated HbA1c (eA1c) < 6.5% (48 mmol/mol) calculated from continuous glucose monitoring values, maintained for at least 3 months without any glucose-lowering pharmacotherapy (10). Testing of HbA1c to document a remission should be performed just prior to an intervention and no sooner than three months after initiation of the intervention or withdrawal of any glucose-lowering pharmacotherapy (10). Although HbA1c is the preferred investigation to define remission, FPG or eA1c needs to be considered in patients in whom HbA1c is altered due to various other diseases.

Expert panel in 2009 had defined 3 types of remission; partial, complete, prolonged (6).

- Partial remission -HbA1c <6.5% without glucose-lowering medications for 1 year
- Complete remission - HbA1c <5.7% without glucose-lowering medications for 1 year
- Prolonged remission – Complete remission lasting more than 5 years

**Therapeutic approaches for remission in T2D**

There are several approaches where remission of T2D had been shown to achieve successfully; low carbohydrate diet, very low calory diet, exercise and bariatric surgery.

***1. Low carbohydrate diet***

This type of carbohydrate restricted diet had been prescribed by clinicians frequently for patients with diabetes before the introduction of exogenous insulin (13). However, with introduction of insulin, the practice has changed and emphasis on low carbohydrate diet (LCD) has been reduced. Definition of LCDs and very low carbohydrate diet is shown in Table 1 (14). Focus in these diets are on macronutrient changes over calory restriction. Protein consumption in these diets are generally unchanged from a standard recommended diet (around 20% of intake), with the remaining energy needs met by fat from either the diet or mobilized body fat stores. Carbohydrate sources are primarily non-starchy vegetables with some nuts, dairy, and limited fruits (15).

Table 1: Definition of low carbohydrate diets

| Definition | Carbohydrate (g/day) | Carbohydrate (% of energy) |
|---|---|---|
| LCD | 20-50g | 6-10% |
| Very LCD | <130g | <26% |

Controlled trials investigating the effectiveness of LCDs are scares and non-controlled heterogenous in design and

outcome. However, a study comparing LCD vs energy restricted low glycemic diet showed 95% of participants could stop diabetic medications compared to 62% of the controls (16). Another study showed diabetes remission in 55% on very low carbohydrate diet compared to 0% in low fat diet (17). Bhanpuri et al had shown that 60% patients on LCD had achieved diabetes remission and significant reduction of medications and insulin in 94% of users (18). At one-year, retention rate was 83%. Similar remission rates (54%) and retention rate (74%) were observed at 2 years in another study by Shai et al. where participants with a mean duration of diabetes of 8.4 years were included. Additionally a weight loss of 10% was also noted (19).

Advantage of LCD is the cost effectiveness, with additional savings from cut down of anti-diabetic medications. Improvements in glycemic control usually appear early and before significant weight loss. LCDs may also have an advantage for long-term weight maintenance. It is also endorsed by ADA and the European Association for the Study of Diabetes (EASD) as an appropriate diet patter for patients with diabetes (20).

Major limitation of LCDs is lack of long-term randomized trials. Follow-up is limited to 2 years. Available studies are heterogenous in study design, inclusion/exclusion criteria and outcome measures. Maintaining LCDs long term would be challenging as for some patients, carbohydrates are addictive.

### *2. Very low-calorie diet (VLCD)*

VLCDs in promoting remission have differences in their study design, including the duration of the intervention (8–20 weeks) and the calorie intake (510–853 kcal/day) (21). However, landmark trials Look AHEAD and DiRECT has provided some answers to the areas of uncertainty. Look AHEAD trial randomized 5145 overweight or obese patients with T2D to an intervention group that received either an intensive lifestyle intervention (ILI) including calorie restriction and increased physical activity or to a control group that included diabetes support and education. Post hoc analysis of this study revealed that at one year, 11.5% of the participants in the ILI group achieved remission (partial or complete); however, remission rates subsequently decreased over time (9.2% at year two and 7.3% at year four). Diabetes Remission Clinical Trial (DiRECT) was planned to overcome the heterogenicity of available studies and expanded over 24 months, enrolling patients with T2D diagnosed within 6 years by the time of enrolment. It comprised of total diet replacement (825–853 kcal/day formula diet for 3–5 months), stepped food reintroduction (2–8 weeks), and structured support for long-term weight loss. According to its results, significant reductions in body weight achieved through VLCD, especially if they are greater than 15 kg compared to baseline weight, are highly predictive of remission in people with diabetes (8).

Taken together, evidence suggests that a LCD is effective in reversing diabetes in the short term up to two years, and its effectiveness was predominantly demonstrated in those with shorter duration since diabetes diagnosis. However, long-term achievement of diabetes remission, adherence to the diet, and weight loss maintenance after the diet remain a challenge.

### *3. Pharmacotherapy*

Sodium-glucose cotransporter 2 inhibitors (SGLT2i) are a novel class of anti-diabetic medications having cardiorenal protection

as well as glycemic effects. It is shown to induce remission of T2D in some studies and postulated mechanism is glucosuria regardless of insulin resistance together with calory loss causing reduction of visceral fat and weight loss. This effect is amplified when other anti-diabetic medications are also used and McInnes et al. showed remission in 24.7% of patients treated with metformin, basal insulin and SLCT2i compared to 16.9% of the group not treated with SGLT2i (22).

Although randomized controlled trials are not available, GLP-1 receptor agonists (GLP-1a) show promising results in weight loss, thus the potential of remission in T2D. Semaglutide showed an average weight reduction of 10 kg together with an average HbA1c of 6.4% with its use (23). Tirzepatide, dual GIP/GLP-1 receptor co-agonist showed that 51.7% of the individuals treated achieved an HbA1c of 5.7% and an average weight loss of 9.5 kg (24).

Other medications like metformin, pioglitazone, anti-obesity drugs have shown varying degrees of weight loss but definitive remission rates of diabetes is not proven yet.

***4. Bariatric surgery***

Around 75% of patients with T2D are overweight or obese (25). Bariatric surgery is aimed at modifying gastrointestinal tract to reduce calory absorption aiming weight loss. Bariatric surgery is indicated in the following;

- For obese patients with a body mass index (BMI) ≥35 kg/m2, regardless of presence, absence, or severity of co-morbidities
- For individuals with metabolic disease and BMI of 30-34.9 kg/m2.
- BMI thresholds should be adjusted in the Asian population such that a BMI ≥25 kg/m2 suggests clinical obesity, and individuals with BMI ≥27.5 kg/m2 should be offered MBS.

It was observed that bariatric surgery leads to remission of diabetes, where exact mechanism is not fully understood to-date.

Several studies showed that in addition to weight loss, an improvement in glucose tolerance is observed which would be mediated by a dramatic improvement in insulin resistance of approximately 50% within one week after surgery (26). In one long-term study, T2DM patients who had undergone bariatric surgery had a reversal rate of over 51% at 12 years, with an average of 35kg weight loss, representing a reduction of 26.9% weight from baseline (27). Long-term outcomes from bariatric surgery of course depend on multiple factors, including type of surgery, patient baseline co-morbidities, patient willingness to engage with lifelong lifestyle change, and the quality of ongoing surveillance. Data suggested that gastric bypass is the most effective at inducing diabetes remission in T2DM patients, followed by sleeve gastrectomy, and then gastric banding (28).

Even after remission following surgical intervention, T2DM patients can still relapse. The prospective Swedish Obese Subjects study reported remission rates of T2DM at 2, 10 and 15 years of follow-up as 72.3%, 38.1% and 30.4%, respectively indicating a proportion of patients relapsing after initial remission (29). Thus, larger prospective randomized studies would be needed to identify the optimal treatment strategies for post-bariatric weight regain and relapse of T2DM with residual or recurrent metabolic disease.

**T2D remission in real world**

Remission rates in community setting are low; 7-year cumulative incidence (in the

absence of bariatric surgery) of partial, complete or prolonged remission was found to be 1.47% (1.40%-1.54%), 0.14% (0.12%-0.16%) and 0.007% (0.003%-0.020%) respectively (30). This is due to several factors.

1. Challenge of maintaining long-term weight loss and lifestyle change.
2. Less awareness among medical community and patients regarding possibility of remission of T2D.
3. Lack of community-based support systems for education, motivation and follow-up of lifestyle interventions
4. Not getting due emphasis about achieving remission in T2D in local/ reginal guidelines
5. Interventions targeting remission of T2D not being the first line of standard of care in management of T2D in available guidelines

Thus, strategic approach at community level should be implemented to overcome the above challenges.

## Conclusion

The increasing incidence of diabetes is an emerging global concern. However, there is evidence that in the initial stages after diagnosis, significant weight loss with surgical or dietary approaches can induce remission of diabetes at a rate greater than 50%. At the same time, new medications, such as SGLT2 inhibitors and GLP-1 analogues or their combination, have a secondary weight loss effect, which can lead several patients to remission. However, maintaining long-term remission requires continuous medical supervision and support from healthcare providers through a personalized approach. Considering that remission has only recently emerged as a therapeutic goal in T2D, awareness among medical community, empowering remission strategies through incorporation in to standard care of T2D and conducting more research to review un-answered areas in remission of T2D is of utmost importance.

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# AN AUDIT ON THE IMPACT OF AGE-ADJUSTED THYROID STIMULATING HORMONE REFERENCE INTERVAL FOR THE ELDERLY, ON CLASSIFYING THYROID STATUS IN PATIENTS AGED OVER 60 YEARS

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## Abstract

**Introduction:** Thyroid Stimulating Hormone (TSH) is the first-line thyroid function test to assess thyroid status. Most laboratories use a uniform reference interval (RI) for TSH in adults. In the elderly, due to the reduced metabolic rate, minor TSH elevations may not be pathological. Therefore, common adult TSH RI can lead to over diagnosis of sub clinical hypothyroidism (SCH) in elderly people, leading to unnecessary treatment.

**Materials and Method :**Data was collected from the TSH requests of the biochemistry laboratory, Apeksha Hospital, Maharagama from March to May 2021, in patients aged >60 years, in whom thyroid function tests (TFTs) were requested for the diagnosis of thyroid disease. Patients with thyroid malignancy and on treatment for thyroid disease were excluded.
Common RI of TSH for the adults and the age-adjusted TSH RI for the elderly from a recognized site were used for the interpretation. Percentage of similar thyroid status by both RIs and the percentage of re-classified thyroid status by age-adjusted RI were calculated.

**Results:** 17.1% of the TSH results indicated SCH according to common adult TSH RI, which is reduced to 9% when interpreted with age-adjusted TSH RI. 88.4% of TSH results showed similar thyroid status by both TSH RIs.8.1% of the TSH results which indicated SCH by common adult RI were re-classified as normal with the use of age-adjusted TSH RI for the elderly.

**Conclusions:** Using a uniform adult RI for TSH in the elderly population (age > 60 years) can lead to over diagnosis of SCH in the elderly leading to unnecessary treatment. Age-specific TSH RIs should be used for people aged > 60 years.

**Key words**- Thyroid stimulating hormone, Subclinical hypothyroidism, age-adjusted reference interval

## Introduction

TSH is the first-line TFT to assess thyroid status. Most laboratories use a uniform RI for TSH in adults. In the elderly, due to the reduced metabolic rate, thyroxin requirement is reduced. Minor TSH elevations may be a normal phenomenon in the elderly. [(1)]
Therefore interpretation with common adult TSH RI can lead to over diagnosis of SCH in elderly people leading to unnecessary treatment. American thyroid association guidelines on the management of thyroid disease suggest raising the target TSH in people over 70 years (up to 4- 6mIU/L0. [(2)]

## Objectives

- Analysis of the TSH reports of biochemistry laboratory, Apeksha Hospital Maharagama from March to May 2021
- Classification of thyroid status of patients using common adult TSH RI verses age adjusted RI for the elderly.
- Evaluate the impact of age adjusted TSH RI on classification of the thyroid status of the elderly.

## Methods

**Study setting:** Biochemistry laboratory, Apeksha Hospital, Maharagama

**Study population:** TSH results of the biochemistry laboratory from March 2021 to May 2021, in patients aged >60 years, in whom TFTs were requested for the diagnosis/screening of thyroid disease.
Patients who are <60 years of age, on treatment for thyroid disease, critically ill patients and patients with thyroid malignancy were excluded.

## Audit Standard [3]

Common RI of TSH for the adults :0.4 – 4.2 mU/L

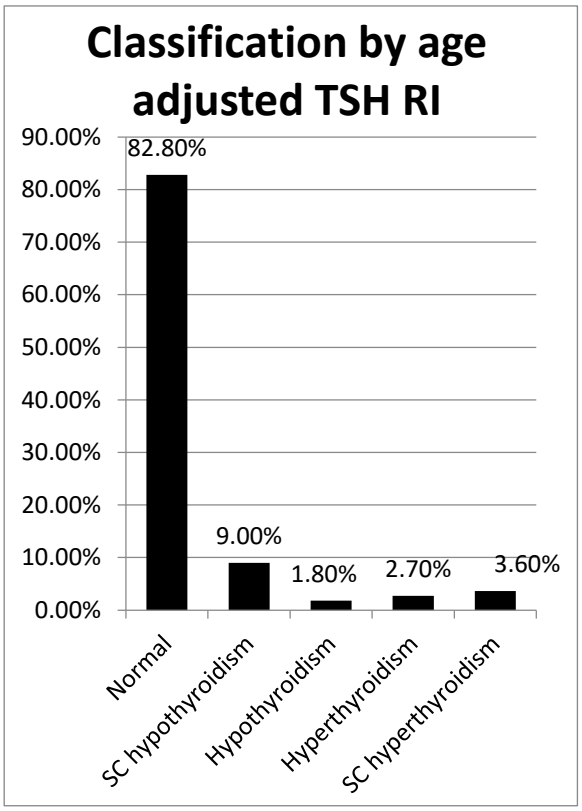
Age-adjusted TSH RI for the elderly:

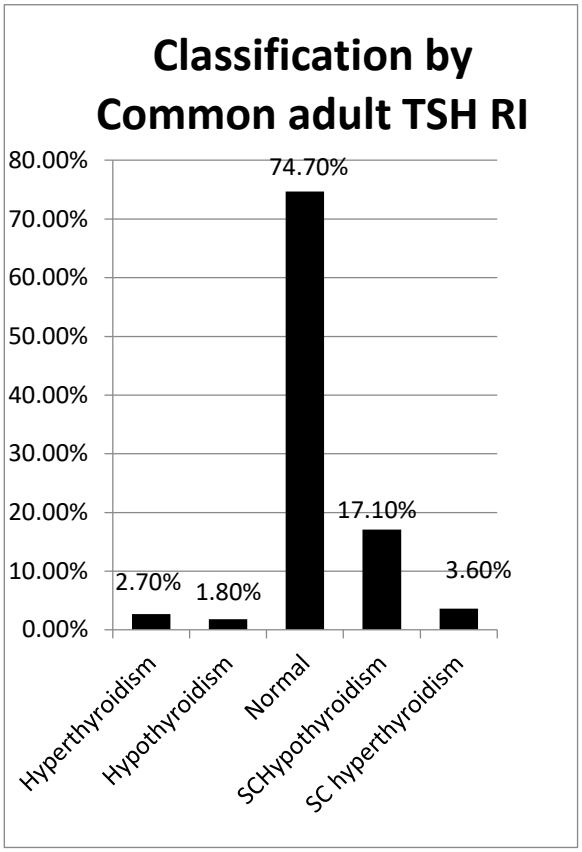
60 -79 yrs.: 0.4 – 5.8mIU/L,
>80 yrs.: 0.4 –. 6.7mIU/L

## Results

Altogether 112 TSH results were included in the study, including 30 males and 81 females. Out of 112 TSH results, 17.1% indicated SCH according to common adult TSH RI, which is reduced to 9.0% when interpreted with age adjusted TSH RI.
Though the majority (91.9%) of TSH results showed similar thyroid status by both TSH RIs, 9.0% of the TSH results which indicated SCH by common adult TSH RI, were re-classified as normal, with the use of age adjusted TSH RI.

| Classification of thyroid status | By Common adult TSH RI | By age specific TSH RI |
|---|---|---|
| Normal | 83 (74.7%) | 92 (82.8%) |
| Hypothyroidism | 2 (1.8%) | 2 (1.8%) |
| Hyperthyroidism | 3 (2.7%) | 3 (2.7%) |
| SC Hypothyroidism | 19 (17.1%) | 10 (9.0%) |
| SC Hyperthyroidism | 4 (3.6%) | 4 (3.6%) |



## Conclusion

Using a uniform adult RI for TSH in the elderly population (age > 60 years) can lead to over diagnosis of SCH in the elderly leading to unnecessary treatment.
Age-specific TSH RIs should be used for people aged > 60 years.

## Recommendation

The Impact of age adjusted TSH RI on classifying the thyroid status of the elderly should be further evaluated in a general hospital rather than in a cancer centre, using a larger study population preferably by a prospective study, in order to include the age adjusted reference range for the elderly in to the TSH report.

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# FREQUENCY IN DIAGNOSING SIGNIFICANT ENDOMETRIAL PATHOLOGIES USING PIPELLE ASPIRATION BIOPSY IN PATIENTS WITH ABNORMAL UTERINE BLEEDING: A RETROSPECTIVE STUDY CONDUCTED AT DISTRICT GENERAL HOSPITAL, NUWARA ELIYA

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## Abstract

**Introduction:** Patients with endometrial pathologies are commonly presented with abnormal uterine bleeding. Endometrial sampling is an effective and simple 1st line diagnostic step in an outpatient setup. Pipelle aspiration biopsy is ideal for obtaining endometrial samples in an outpatient department which is more convenient, having a high overall accuracy and sensitivity in diagnosing endometrial pathologies.

**Objective:** To determine the frequency of diagnosing endometrial pathologies using Pipelle aspiration biopsy.

**Materials and Method:** All the pipelle aspiration specimens received during a period of five months from 01st June to 31st October, 2022 were included in the study. Samples with request forms having flaws were excluded. The patients' age, clinical presentation and the histopathological findings were obtained. All cases were stratified into age groups 21-40 years(reproductive), 41-50 years(perimenapausal) and >50 years(postmenapausal). The histological findings were categorized as normal pattern (NP), exogenous hormonal effects (EHE), non-phasic endometrium (NPE), pathological endometrium (PE) and non-diagnostic samples (NDS).

**Results:** A total of 47 pipelle aspiration specimens were analysed. The age range of the patients was 21–80 years (reproductive - 2, perimenapausal- 19, postmenapausal- 26). Normal pattern was found in 18%, EHE- 2%, NPE- 15% and PE- 18%. There were 47% of non-diagnostic samples despite the clinical presentation. Only 53% of samples were adequate for assessment.

**Conclusion:** Percentage of obtaining adequate endometrial tissue for assessment in pipelle aspiration biopsy in DGH, Nuwara Eliya is low when compared to the gold standards.

**Key words:** endometrial pathologies, frequency, pipelle aspiration biopsy.

## Introduction

Abnormal uterine bleeding (AUB) is defined as bleeding from the uterine corpus that is abnormal in regularity, volume, frequency, or duration and occurs in the absence of pregnancy [1].

It is a significant entity affecting 14–25% of women of reproductive age and may have a significant impact on their physical, social, emotional and material quality of life [2].

It is a frequent symptom in perimenopausal women which negatively affects the woman's quality of life and psychological well-being [3].

Aetiology of AUB can be classified as structural and non-structural causes and according to the International Federation of Gynaecology and Obstetrics (FIGO) classification system, the causes of AUB includes endometrial polyp, adenomyosis, leiomyoma, malignancy and hyperplasia, coagulopathy, ovulatory dysfunction, endometrial pathology, iatrogenic cause, and those causes not yet classified (PALM-COEIN) [4].

Endometrial hyperplasia (EH) is often suspected in women presenting with abnormal uterine bleeding. Trans-vaginal ultrasound plays a role in diagnosing endometrial pathology in pre- and postmenopausal women [5].

However, confirmation of diagnosis requires histological analysis of endometrial tissue specimens obtained either by using outpatient suction devices designed to blindly abrade and/or aspirate endometrial tissue from the uterine cavity or by inpatient endometrial sampling, such as dilatation and curettage performed under general anaesthesia [5].

Pipelle biopsy is ideal for obtaining endometrial sample in outpatient department. It samples only 4% of the endometrial surface and has sensitivity up to 97% [6] , [7]. It is widely used in women with abnormal uterine bleeding or postmenopausal bleeding. The method became very useful due to its simplicity as well as high sensitivity in detecting carcinoma [8]. In one of the early studies, the technique was described as adequate for analysis in 97% of patients [8]. Pipelle device has become superior enabling both outpatients sampling and decreasing the cost of the service [8]. The Pipelle is the best device in both postmenopausal and premenopausal women, with detection rates of 99.6% and 91%, respectively in diagnosing EH and carcinoma. Pipelle has a sensitivity of 81% and specificity of more than 98% in detecting atypical EH [8].

Failure to obtain adequate samples for histological examination is one of the major issues associated with the Pipelle procedure. Different factors have an impact on the procedure success: patient related-factors like prior cervical procedures, genital anomalies, obesity; and provider related factors like the inability to access endometrium, inadequate sample, physicians' experience, etc. A more recent study revealed that pipelle failed in 22.89%, where in 17.39% it happened due to inability to reach the endometrium, in 80.43% were inadequate samples, and in 2.18% was due to unknown reasons [8].

## Materials and Method

This study was a retrospective, cross-sectional study conducted at the Department of Pathology, District General Hospital, Nuwara Eliya for a period of five

months from 01st June to 31st October 2022

All the pipelle aspiration specimens received during this period were included in the study. Samples with request forms having flaws were excluded. These cases were selected from the departmental data base. The request forms and the haematoxylin and eosin stained slides of all the cases were retrieved.

The patients' age, clinical presentation, ultrasound scan findings and the histopathological findings were obtained.

All cases were stratified into age groups 21-40 years (reproductive), 41-50 years (perimenopausal) and >50 years (postmenopausal).

Abnormal uterine bleeding is considered as the presenting complain which is further divided in to intermenstrual bleeding (IMB), heavy menstrual bleeding (HMB), perimenapausal bleeding, post-menopausal bleeding (PMB) and other complaints including vaginal discharges, vaginal lumps, adnexal masses, dysmenorrhea, invasive breast carcinoma and chemotherapy treatment.

Macroscopic findings were classified as large, moderate and scanty amounts.

All the slides were reviewed by the Acting Consultant Histopathologist, DGH, Nuwara Eliya

The histological findings were classified as follows.

1. Normal pattern (NP) – This includes proliferative phase endometrium and secretory phase endometrium.
2. Exogenous hormonal effects (EHE)
3. Non-phasic endometrium (NPE)
4. Pathological endometrium (PE) – This includes benign endometrial polyp (BEP), acute inflammation (AI) / Endometritis, disordered proliferative endometrium (DPE), endometrial hyperplasia (EH) and carcinoma (CA)
5. Non-diagnostic samples (NDS) – This includes samples having mucous only, blood only, no endometrial tissue, ectocervical tissue only and samples inadequate for assessment.

Overall frequency of any kind of these histological findings was grouped according to the presentation and age group.

**Results**

A total of 54 pipelle aspiration samples submitted for histological diagnosis during the period of five months were analysed. The study sample is 47 and 7 samples were excluded from the study due to having flaws in the request forms.

The age range of the patients was 21–80 years. Most of them were in postmenapausal (>50 years) age group.

**Table 1.** Distribution of cases in different age groups

| **Age groups (Years)** | **Number of cases** |
|---|---|
| **Reproductive (21-40)** | 4% (2) |
| **Perimenapausal (41-50)** | 41% (19) |
| **Postmenapausal (> 50)** | 55% (26) |

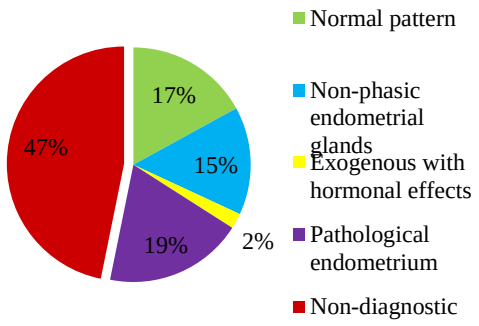
34% were presented with PMB. HMB and perimenapausal bleeding were equally presented and it was 23%. IMB is the presenting complaint of 4%. 15% has had other symptoms.

94% (44/47) of patients in our study have undergone pipelle biopsy for the first time

and other 4% (3/47) were repeat biopsies. Majority of the patients were having thick endometrium and a bulky uterus ultrasonically. There were no large samples received for the assessment. 26% of macroscopically moderate amount samples were analysed. Other 74% were scanty samples.

In our study 47% (22/47) were non-diagnostic samples. Only 53% of samples were having any kind of diagnosis.

**Figure 1. Frequency of histological findings**



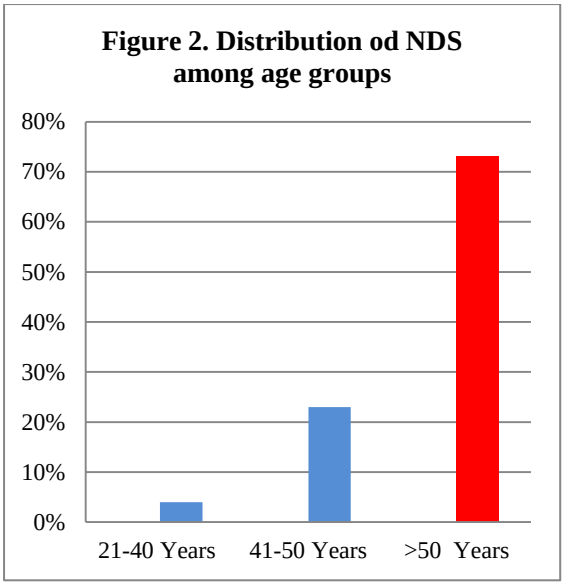

Normal pattern was found in 17% cases; Proliferative phase endometrium - 9% and Secretory phase endometrium - 9%. 2% were diagnosed as EHE. NPE was the diagnosis in 15%.

Total of 19% were having a pathological endometrium (PE), including BEP - 4% , AI/ Endometritis- 4%, DPE - 4%, EH - 4% and CA -2%.

Out of these NDS, 73% (16/22) were from postmenapausal (>50 years) age group where majority were presented with PMB and 23% (5/22) were from perimenapausal (41-50 years) age group and only 4% (1/22) were from reproductive (21-40 years) age group.

**Table 2.** Distribution of histological findings in AUB

| **Histopathological finding** | **Frequency** |
|---|---|
| **Non-diagnostic** | **47%** |
| **Normal pattern** | **17%** |
| Proliferative phase endometrium | 9% |
| Secretory phase endometrium | 9% |
| **Pathological endometrium** | **19%** |
| Benign endometrial polyp | 4% |
| Acute inflammation/ Endometritis | 4% |
| Disordered proliferative endometrium | 4% |
| Endometrial hyperplasia | 4% |
| Carcinoma | 2% |
| **Non-phasic endometrium** | **15%** |
| **Exogenous hormonal effects** | **2%** |




## Discussion

Abnormal uterine bleeding is the most common presenting symptom of women presented to outpatient department who need further investigations to exclude structural causes of AUB. In such occasions, endometrial sampling can be the most effective simple diagnostic method. Pipelle biopsy is ideal for obtaining endometrial sample in outpatient department. It samples only 4% of the endometrial surface and has sensitivity up to 97% in initial diagnosis of endometrial pathologies especially endometrial hyperplasia. It is very cost effective in diagnosis and early treatment [(6).]

In our study most of the pipelle biopsy samples were from postmenapausal (>50 years) age group (55%) and comparatively less amount were getting from perimenapausal (41- 50 years) age group (41%), though perimenapausal (41-50 years) group was the most frequently investigated in other studies who presented with AUB. They are the most vulnerable groups for endometrial hyperplasia. The incidence of endometrial hyperplasia is estimated to be at least three times higher than endometrial cancer and if left untreated it can progress to cancer [(5).] According to our study, only 53% were having a diagnosis.

Pathological endometrium (PE) was diagnosed in 19% and normal pattern (NP) was diagnosed in 17% in our study. The next common histological finding was NPE and it was 15%. Only 4% were diagnosed as EH and incidence of CA was 2% in the index study.

Nearly half (47%) of the pipelle aspiration samples received to department of pathology, district general hospital, Nuwara Eliya were concluded as non-diagnostic samples though most research found pipelle aspiration biopsy having high sensitivity and it is adequate for analysis in 97% of patients [(8).]

The incident of getting a NDS is 47% in district general hospital, Nuwara Eliya. Out of this, 73% were from postmenapausal (>50 years) age group women and 23% were from postmenapausal (41-50 years) age group women.

Samples having mucous or blood only, samples not having endometrial tissue and samples inadequate for assessment were considered as NDS. Samples having only ectocervical tissues were also included in NDS category. Only 26%of samples were having moderate amount of tissue microscopically while 74% of samples having scant amount of tissue. Pipelle aspiration biopsy has a high accuracy in diagnosing endometrial cancers when an adequate specimen is obtained [(9)]. The inappropriate techniques, unexperienced hands, sample transportation errors may

have some contribution for this higher frequency of NDS.

## Conclusion

The frequency of getting non diagnostic samples of pipelle aspiration biopsy is higher than the diagnostic samples. The frequency of diagnosing endometrial hyperplasia and endometrial cancers in high risk population (perimenapausal and postmenapausal women) is still in the lower side. So there is a large discrepancy in frequency of diagnosing endometrial pathology via pipelle aspiration biopsy in district general hospital, Nuwara Eliya. The reasons for this should be re-audited and the proper implementations should be taken aiming to diagnosing the patients with endometrial pathologies. However, repeat biopsy via dilatation and curettage or hysteroscopy is crucial when the initial histopathological assessment is doubtful compared to patients' clinico-radiological presentation to avoid misdiagnosis.

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# STUDY ON COMPLICATION SCREENING PRACTICES IN PATIENTS WITH DIABETES MELLITUS AT OUT-PATIENT DEPARTMENT AT RURAL HOSPITAL SETTINGS: A SINGLE CENTRE CROSS SECTIONAL STUDY

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[1]District General Hospital, Mullaitivu

## Abstract

**Background**: Diabetes mellitus (DM) is a major health problem in family practice causing multiple micro- and macro-vascular complications, thus prevention of such would be the main aim of the physician. Lack of proper assessment can hasten the complications and a meticulous screening system is a prerequisite in every diabetic patient's evaluation.

**Aim:** The aim of the study was to assess the pattern of end organ screening for diabetes related complications in patients with type 2 DM for early detection of complications by physicians in an outpatient setting in a rural hospital.

**Results:** Of 104 study participants (Male 63.5%; Age 61years ± 7.6SD), 100% had blood pressure monitoring as per recommendation while retinopathy, nephropathy and neuropathy assessment were 26.4%, 64.4% and 0% respectively. Cardiovascular risk assessment was done in 20.2% while none had advice on comprehensive foot care.

**Conclusion:** Screening for diabetes related complications needs more emphasis and attention at the given setting to meet international standards.

## Introduction

As per statistics of International Diabetes Federation (IDF) 2022, Sri Lanka has 1417600 patients diagnosed with diabetes across all age groups. Data also shows a 9.8% increase in prevalence of diabetes.[1] Diabetes and its complications impose a significant economic impact for individuals and their families, as well as the country's health care.[5]

Poor glycaemic control can hasten the micro and macro vascular changes, predisposing to serious complications.[2-4] Longer the duration of the disease, incidence of complications and severity increases.

Primary care physicians are the first point of contact and sometimes the only treatment provider for majority of the patients, especially in a rural area like Mullaitivu. Hence, there is a need of awareness among these treating physicians on systemic screening to identify the complications and take quick preventive and therapeutic interventions. American Diabetes Association (ADA) guidelines strongly

recommend complication screening and surveillance. [2-4]

## Objectives

1. To observe if screening methods for macrovascular and microvascular complications are used routinely in diabetic patient care
2. To assess the adherence of the physician to the diabetic care guideline
3. To look for lacuna in the screening for preventable complications

## Methods

A cross sectional study was conducted in District General Hospital Mullaitivu during the month of November 2022 at outpatient medical clinic. Formal approval was obtained from the hospital authority before commencement of the audit. A total of 104 patients, selected on convenient sampling method, who were on follow up for diabetes, were assessed with their informed verbal consent. Patients were interviewed on whether they underwent screening for micro and macro vascular complications according to ADA guidelines (American Diabetes Association Professional Practice Committee, 2022a) (American Diabetes Association Professional Practice Committee, 2022b) (American Diabetes Association Professional Practice Committee, 2022c).

Inclusion criteria

All diabetic patients on follow up for type 2 DM presenting to the outpatient medical clinic for routine visit.

Exclusion criteria

- Type 1 diabetes
- Gestational diabetes
- Less than 1 year following diagnosis of diabetes

Patients on follow up for diabetes were assessed on the following parameters;

- Blood pressure monitoring at each visit
- Ophthalmological examination at the time of diagnosis and annually
- Cardiovascular risk assessment with 12 lead ECG annually and lipid profile at the time of first contact and once every 6 months,
- Nephropathy screening with annual urine albumin/creatinine (ACR) ratio or UFR if there is gross proteinuria in the absence of urine tract infection
- Assessing for distal peripheral neuropathy at diagnosis and annually by clinical assessment and tuning fork test
- Providing comprehensive foot care at diagnosis and annually by monofilament test and doppler pulse study

Patient information were collected from clinic records and from the patient using a questionnaire a single physician and recorded.

## Results

Of 104 patients screened, 63.5% were males and the mean age was 61 years (± 7.6SD). Frequency of clinic visits are once a month per year. Measurement of blood pressure was done in all the participants (100%) at

each clinic visit.54.8% subjects had a lipid profile done during the last six months. Annual cardiovascular risk assessment had been performed in 20.2% of patients. 64.4% had assessment of microalbuminuria or urine ACR as per recommendations. Only 26.4% had undergone retinopathy screening with in last 12 months while30.8% has never had a retinopathy screening before.17.3% had already developed retinopathy which was only detected after patient developed poor vision.

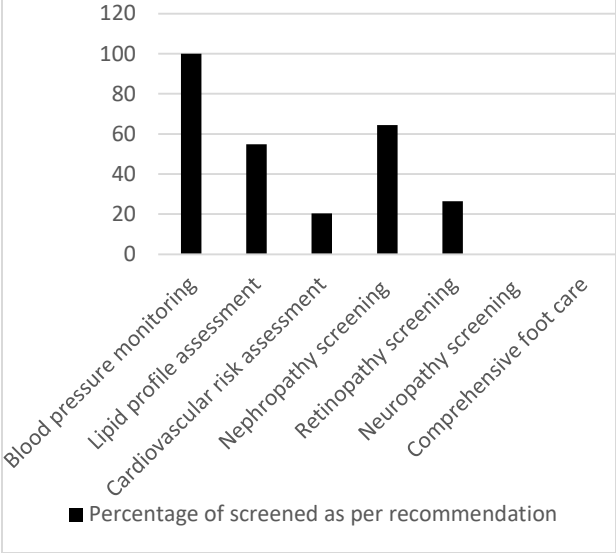


Figure 1: Percentage of patients screened for micro and macrovascular complications

None of the subjects were screened for diabetic neuropathy. Advice on comprehensive foot care was also not a regular practice among physicians.

## Conclusion

End-organ screening for diabetes related complications had been performed only on some patients as per standard recommendations at the given OPD set-up. Evaluations which consume more time like foot assessment are less commonly performed on patients while routine assessment like blood pressure monitoring are done in almost all patients with diabetes.

## Discussion

Our study showed that a significant percentage of patients have not had the full screening of macro- and microvascular complications and key assessment components of diabetic complication screening had being missed by the treating doctors. Reasons for the non-assessment of certain important criteria would include;

- Less time spent per patient due to large number of patients seen by a physician in a day.
- Patients refusing investigations as they are asymptomatic
- Limited availability of investigations at peripheral hospitals
- Lack of dedicated diabetic clinic in the OPD with trained diabetic care nursing officer
- Lack of knowledge of the physician of standard protocol for systematic follow up

Limitations of the study

This is a baseline clinical study and subsequent clinical audits must be carried out to assess the progress in the quality of care following an appropriate intervention. Moreover, the current study may not be the complete reflection of the assessment of all the parameters since the documentation of few essential data may be missed during consultations in a busy OPD settings.

**Recommendations**

1. To educate doctors, patients and caregivers "to look beyond sugar control", importance of end-organ screening for diabetes related complications.
2. To make patients aware on annual ophthalmic examination, microalbuminuria, or urine ACR testing, complete foot examination during the routine healthcare visit
3. To establish national and institutional Standard Operating Protocols (SOPs) on diabetes related complication screening and educate health care professionals on them.
4. Special training programs or continued medical education for healthcare professionals, especially working in the peripheral/ rural healthcare institutions to update on the importance of annual screening for diabetes.
5. To establish dedicated diabetic treatment clinics where only patients with diabetes are seen and to allocate a diabetes educator nursing officer to each such clinic.
6. To standardise the management and follow up goals ensuring uniformity in terms of diabetic care and its associated complications.
7. To conduct periodic audits to assess the change in quality of care.

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# PREVALENCE OF NON-ALCOHOLIC FATTY LIVER DISEASE ACCORDING TO THE NAFLD SCORE AND ASSOCIATED RISK FACTORS AMONG PATIENTS ADMITTING TO NATIONAL HOSPITAL KANDY

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## Abstract

**Background:** Non-Alcoholic Fatty Liver Disease (NAFLD) has been recognized worldwide and is considered a leading cause of cirrhosis. An only a limited number of researches have been done about the Asian population. Most of the available studies have been based on invasive methods like liver biopsies. NAFLD Score has been developed as a non-invasive method of identifying the level of liver fibrosis giving more advantage over the invasive methods.

**Objectives:** The objective of this study was to determine the prevalence of Non-Alcoholic Fatty liver disease among patients admitted to National Hospital Kandy according to the NAFLD Score and to assess the associated common risk factors. And to identify the importance of screening the patient with metabolic risk factors for NAFLD.

**Methodology:** The study was a descriptive, cross-sectional study and the study period was from 15 March 2018 to 15 May 2018. The study setting was medical units at National Hospital Kandy. During this study, the NAFLD fibrosis score was categorized into three major categories, no fibrosis, intermediate fibrosis, and advanced fibrosis in the liver respectively.

**Results:** This study consisted of 200 patients with the age between 30 years and 70 years. The majority of the study population was female (n=104, 52%). The mean age of the study population was 54 years (SD= 12 years). The majority were in the 'Over Weight' category (50.5%). The mean value of body mass index in the study group was 24.95 Kg/m2 (SD=4.68).
Among male, 58.3% had no fibrosis and less than 5% had advanced liver fibrosis. No fibrosis in females showed 56.7% while advanced liver fibrosis was less than 5%. The association between gender and fibrosis status was not significant (p=0.592).

The majority of the study population did not have diabetes mellitus (n=131) and the majority of the diabetic mellitus group had intermediate fibrosis (63.7%). The association between diabetes mellitus status and liver fibrosis status was highly significant ($p < 0.001$). The hypertension prevalence of the study population was 48% and out of that, 50% had intermediate fibrosis status. The association between hypertension status and liver fibrosis status is significant ($p < 0.001$). When the study categorized into low, normal, overweight, and obese categories depend on their BMI, the majority of the study population found to be in

the overweight category, which was 50.5%. The majority of all four categories of the study populations had no fibrosis but sixteen percent of the obese group had advanced liver fibrosis
**Conclusion:** This study provides very impotent information about the local prevalence rates for NAFLD and related liver fibrosis status in our local population and thereby the clinical and economic burden of NAFLD in a patient with metabolic risk factors about demographical variations of the Sri Lankan population.

**Key words:** Non- Alcoholic Fatty Liver Disease, Liver fibrosis, Non-Communicable Diseases, Sri Lanka

## INTRODUCTION

Nonalcoholic fatty liver disease (NAFLD) has become a major health problem worldwide because it tended to evolve into cirrhosis and hepatocellular carcinoma (HCC) [1]. The nonalcoholic fatty liver disease represents a spectrum of liver diseases including the simple fatty infiltration of the liver parenchyma (steatosis), fatty infiltration and inflammation NASH, and cirrhosis, in the absence of immoderate alcohol consumption [2,3]. Epidemiological studies from different countries have indicated liver fibrosis is the best predictor of liver-related and overall mortality in patients with NAFLD [4]. Diagnosis of steatosis and liver fibrosis can be done using laboratory tests, different imaging methods, and liver biopsy. The clinical examination does not help for the early diagnosis of liver changes related to NAFLD, as most of the patients in the early stage of fatty liver disease have no clinical signs or symptoms. Imaging studies used to identify changes in the liver-related to NAFLD include conventional ultrasound, as well as ultrasound-based methods known as ultrasound elastography (fibro scan). Magnetic resonance elastography (MRE) can also be used for the analysis of liver fibrosis. Liver biopsy is still considered the gold standard for the diagnosis of fatty liver disease [5]. However, liver biopsy is not a suitable test for screening in clinically in significant fibrosis as it is an invasive investigation and also it is a condition that affects at least 30% of adult people.

The prevalence of NAFLD among Sri Lankan adults is rising. In a large population-based study from urban Sri Lanka, among 2,985 adults (aged 35-64 years), 974 (32.6%) were found to have NAFLD on ultrasonography [6]. The presence of NAFLD has been identified as having strongly associated with the key medical conditions of metabolic syndrome. In a similar study, performed in a rural tea-cultivating, physically active, low-income population of adults, the prevalence rate was expectably lower than the urban figures, but still significant at 18% [7]. The urban cohort, when followed up for 7 years and carried out the ultrasound scan of the liver again, which has been showing their prevalence of NAFLD had increased remarkably to nearly 66% in this new aging (42-71 years) population [8]. The annual incidence of NAFLD in this population was 6.6%.

During the last decade, various noninvasive blood-based and imaging-

based biomarkers have been developed and used for the assessment of the level of liver fibrosis in patients with NAFLD [9,10]. Simple blood-based scores including routine parameters can be easily obtained and these tests are suitable even in the primary care setting [11]. Especially in low resource countries like Sri Lanka, even arranging a simple imaging study like liver ultrasound scanning is not practical. In this type of medical setup, the NAFLD fibrosis score (NFS) can be easily calculated using simple parameters with the aid of online calculators. The NAFLD score has shown good performance for detecting advanced fibrosis and cirrhosis [12].

Though the study-based evidence related to NAFLD is available, most of these studies have been carried out among the European population. The number of studies about Asians is limited and the relevance of the application of NAFLD score to the Sri Lankan population has not been adequately evaluated.

This study aimed to identify the level of liver fibrosis by calculating the NAFLD score in the hospital admitting patients considering as a sample from the draining population to the National Hospital Kandy.

## OBJECTIVES

The objective of this study was to determine the prevalence of Non-Alcoholic Fatty liver disease among hospital admitting patients in National Hospital Kandy according to the NAFLD Score and to assess the associated common risk factors. And to understand the importance of screening patient with metabolic risk factors for NAFLD to minimize the related morbidity and mortality.

## METHOD

This study was design as a descriptive cross-sectional study and the study was carried out in medical units at National Hospital Kandy. The study was carried out from 15 of March 2018 to 15 of May 2018. Study population was the patients who were admitted to the above units with age between 30 years to 70 years.

The following patients were excluded, patient who consumes alcohol exceeding safe limit (12 U per week), the patients who are admitting with acute febrile illnesses, patient who can develop high transaminases due to other conditions (i.e. poisoning, snake bite, acute coronary syndrome, rhabdomyolysis, critically ill patients. etc.)
Sample size was calculated as follows,

$$\text{Sample size} = \frac{Z^2\, p \times (1-p)}{d^2}$$

(Lwanga and Lemeshow)

Z=Z value
p=percentage picking a choice
d = desire precision

[Prevalence of Non-Alcoholic Fatty liver disease among hospital admitting patients as 32.6% and the desire precision assumed as 7%. If the non-responsive rate is 10%, the final sample size required was 189].

Every patient selected consecutively after considering inclusion and exclusion criteria till minimum sample size achieved. During this study the NAFLD fibrosis score was categorized as into three major categories as no fibrosis, intermediate fibrosis and advanced fibrosis in the liver. The NAFLD fibrosis score was calculated for each individual who were participated for this study using the NAFLD score calculate in the QXMD smart application.

The facts which need to calculate above score are the age, BMI, diabetic state, transaminases, platelet count and the serum albumin level.

The inward patient who fulfilled the inclusion criteria were individually educated about the study and written consent was obtained using the consent form prepared in three main languages after getting the approval from the ethical clearance committee.

**Ethical considerations**

Ethical clearance was obtained from the Ethics Review Committee of National Hospital (NH) Kandy. Patient autonomy was respected and informed written consent was obtained prior to collection of data. The principal investigator described the importance of the study.

**Data analysis**

Data analysis was carried out using the appropriate statistical methods in the Statistical Package for Social Sciences (SPSS) version 23.0.

**RESULTS**

The results were obtained after analyzing the data collected by interviewing a total of 200 patients' ages between 30 to 70 years admitted to medical unit, NH Kandy. Response rate was 100%. Majority of the study population was Female (n=104, 52%). Mean age of the study population was 54 years (SD= 12 years). Out of all respondents, Mean Height and Weight of the study population were 156.6 cm (SD= 7.3 cm) and 61 Kg (SD= 11Kg) respectively.

Table 1- Distribution of BMI Categories frequency and percentage

| **BMI Categories** | **N** | **%** |
|---|---|---|
| Low BMI | 13 | 6.5 |
| Normal BMI | 61 | 30.5 |
| Over Weight | 101 | 50.5 |
| Obese | 25 | 12.5 |
| **Total** | 200 | 100 |

Of the respondent, majority were in 'Over Weight' category (50.5%). Mean value of BMI in the study group was 24.95 $Kg/m^2$ and SD was 4.68.

**Description of NAFLD Score of the study population**

NAFLD Score is categorized as No Fibrosis, Intermediate Fibrosis and Advanced Liver Fibrosis. Majority of the participant (57.5%) was belonging to No Fibrosis group.

Table 2- Distribution of Fibrosis Status frequency and percentage

| NAFLD Score | N | % |
|---|---|---|
| No Fibrosis | 115 | 57.5 |
| Intermediate Fibrosis | 79 | 39.5 |
| Advanced Liver Fibrosis | 6 | 3.0 |
| **Total** | 200 | 100 |

**Association of socio demographic characteristics with NAFLD Score**

Table 3- Association between gender and NAFLD Score

| | No Fibrosis | | Intermediate Fibrosis | | Advanced Liver Fibrosis | | Total | |
|---|---|---|---|---|---|---|---|---|
| **Gender** | **N** | **%** | **N** | **%** | **N** | **%** | **N** | **%** |
| Male | 56 | 58.3 | 36 | 37.5 | 4 | 4.1 | 96 | 100 |
| Female | 59 | 56.7 | 43 | 41.3 | 2 | 1.9 | 104 | 100 |

*$X^2$ value= 1.047, df=2, p=0.592*

Among male, 58.3% had no fibrosis and less than 5% had advanced liver fibrosis. In female, no fibrosis is found to be 56.7% while advanced liver fibrosis was also less than 5%. The association between gender and fibrosis status is not significant (p=0.592).

**Association of Diseases status with NAFLD Score**

Table 4- Association between Diabetes Mellitus and NAFLD Score

| | No Fibrosis | | Intermediate Fibrosis | | Advanced Liver Fibrosis | | Total | |
|---|---|---|---|---|---|---|---|---|
| **DM** | **N** | **%** | **N** | **%** | **N** | **%** | **N** | **%** |
| Yes | 21 | 30.4 | 44 | 63.7 | 4 | 5.9 | 69 | 100 |
| No | 94 | 71.7 | 35 | 26.7 | 2 | 1.6 | 131 | 100 |

*$X^2$ value= 31.874, df=2, p <0.001*

Majority of the study populations had not having Diabetes Mellitus (n=131). Out of that, 71.7% had No Fibrosis status. But, majority of Diabetic Mellitus group had Intermediate Fibrosis (63.7%). The association between Diabetes Mellitus status and Liver Fibrosis status is highly significant *(p <0.001).*

Table 5. Association between Hypertension and NAFLD Score

| | No Fibrosis | | Intermediate Fibrosis | | Advanced Liver Fibrosis | | Total | |
|---|---|---|---|---|---|---|---|---|
| **Hypertension** | **N** | **%** | **N** | **%** | **N** | **%** | **N** | **%** |
| Yes | 44 | 45.8 | 48 | 50.0 | 4 | 4.2 | 96 | 100 |
| No | 71 | 68.3 | 31 | 29.8 | 2 | 1.9 | 104 | 100 |

*$X^2$ value= 10.361, df=2, p = 0.006*

Hypertension prevalence of the study population was 48% (N=96). Out of that, 50% had Intermediate Fibrosis status. Majority of Non-Hypertension group had No Fibrosis status (68.3%). The association between Hypertension status and Liver Fibrosis status is significant *(p <0.001).*

Table 6- Association between Dyslipidemia and NAFLD Score

| | No Fibrosis | | Intermediate Fibrosis | | Advanced Liver Fibrosis | | Total | |
|---|---|---|---|---|---|---|---|---|
| **DL** | **N** | **%** | **N** | **%** | **N** | **%** | **N** | **%** |
| Yes | 93 | 60.4 | 57 | 37.0 | 4 | 2.6 | 154 | 100 |
| No | 22 | 47.8 | 22 | 47.8 | 2 | 4.4 | 46 | 100 |

*$X^2$ value= 1.383, df=2, p = 0.304*

Dyslipidemia prevalence of the study population was 77% (N=154). Majority had no fibrosis (60.4%) and fifty-seven had intermediate fibrosis. The association between Dyslipidemia and Liver Fibrosis status is not significant *(p <0.304).*

Table 7- Association between BMI and NAFLD Score

| | No Fibrosis | | Intermediate Fibrosis | | Advanced Liver Fibrosis | | Total | |
|---|---|---|---|---|---|---|---|---|
| **DL** | **N** | **%** | **N** | **%** | **N** | **%** | **N** | **%** |
| Low BMI | 12 | 92.3 | 1 | 7.3 | 0 | 0.0 | 13 | 100 |
| Normal | 37 | 60.6 | 23 | 37.7 | 1 | 2.3 | 61 | 100 |
| Overweight | 53 | 52.4 | 47 | 46.5 | 1 | 1.1 | 101 | 100 |
| Obese | 13 | 52.0 | 8 | 32.0 | 4 | 16.0 | 25 | 100 |

*$X^2$ value= 24.631, df=6, p < 0.001.*
*Linear by linear association $X^2$=8.056, df=1, p=0.05*

Majority of all four categories of the study populations had no fibrosis. Sixteen percent (n=4) of obese group had advanced liver fibrosis. The association between BMI status and Liver Fibrosis status is highly significant *(p <0.001).* But There is not significant Linear by liner association in the study.

## DISCUSSION

In this study, the majority of male participants (58.3%) had no fibrosis and only less than 5% had advanced liver fibrosis. Among the female population, 56.7% were in the no fibrosis category, while less than 5% were in the advanced liver fibrosis category. According to these results association between gender and fibrosis status is not significant (p=0.592). Thus, this study did not show a significant association between gender and fibrosis status.

A large proportion of the studied sample of patients did not have Diabetes Mellitus (n=131). Out of them, 71.7% found to have no fibrosis. But, the majority of the Diabetic Mellitus group had intermediate fibrosis (63.7%). The association between Diabetes Mellitus status and liver fibrosis status was highly significant *(p <0.001).*

The prevalence of hypertension in the study population was 48% (N=96). Out of them, 50% had intermediate fibrosis status. The majority of the Non-Hypertension group had no significant fibrosis status (68.3%). The association between hypertension status and liver fibrosis status is significant *(p <0.001).*

The dyslipidemia patient number in the study group was 154 giving the prevalence state of 77%, though the majority had no fibrosis (60.4%) and fifty-seven had intermediate fibrosis. However, the association between dyslipidemia and liver fibrosis status is not significant *(p <0.304).*

When considering the BMI status of the study population (Low, Normal, Overweight, and Obese), the majority of all four BMI categories had no fibrosis. Sixteen percent (n=4) of the obese group had advanced liver fibrosis. The association between BMI status and liver fibrosis status was highly significant *(p <0.001).* But there was no significant linear by linear association in the study.

According to this study, it is clear that the risk of fibrosis is high in people who are having metabolic risk factors (single or multiple).

This study shows that majority of the patient who had Diabetic Mellitus have a very significant intermediate fibrosis score. These findings were compatible with the systematic review and the meta-analysis carried out among 80 studies from 20 countries by Younossi ZM et al [13] and the systematic review and meta-analysis of a population-based observational study by Jarvis H et al [14].

Among the studied population, the group who had hypertension, 50% had intermediate fibrosis score making a highly significant association.

Though 77% of the studied population had dyslipidemia, a majority did not have fibrosis making a statistically in significant association between dyslipidemia and liver fibrosis.

Liver diseases are not giving a priority compared to many other communicable and non-communicable diseases in Sri Lanka. However, from the practicing clinicians' point of view, the liver disease carries a significant health problem related to associated morbidity and mortality. The two most common etiologies for liver disease in Sri Lanka are nonalcoholic fatty liver disease (NAFLD) and alcoholic liver disease. Available research data on the

burden of liver disease in Sri Lanka is sparse [15].

The health cost which needs to manage a patient with cirrhosis is huge and the effect of disease-related complications on an individual patient's quality of life is very much significant. Early identification of vulnerable people is crucial for the proper risk factor control and to arrange the regular follow-up of such patient who is at risk. Due to the above facts, it is a timely necessity to perform a regular risk assessment for NAFLD in the patient who is having metabolic risk factors during our clinical practice. It needs only basic blood investigations to calculate the fibrosis score by using the NAFLD score calculator. As we have done in this study, it can easily introduce into our routine clinical practice. In patients followed up in medical clinics, these blood investigations are performed six-monthly or annually, especially during their clinic follow-up. If we can calculate the NAFLD fibrosis score routinely and identify patients with a high-risk profile, it will assist to make a significant alteration in the medical care we are giving for our patients. Especially this simple calculation helps to give the priority to control the risk factor in a patient taking an individualized approach. Thereby necessary management and testament changes (pharmacological and non-pharmacological) can be arranged for risk patients. Eventually, it can delay the progression into advanced fibrosis and end-stage liver disease and help to minimize the high NAFLD-related morbidity and mortality. Moreover, this score can be used as a tool to identify patients who need liver biopsies (gold stranded test to identify liver fibrosis) which is an invasive procedure not practical to arrange for those who are having a high-risk factor profile for NAFLD. Ultimately, the findings revealed from this study can be implemented into our routine clinical practice to ameliorate patient care in the future.

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# ASSESSMENT OF KNOWLEDGE AND ATTITUDES WITH REGARD TO PAIN MANAGEMENT AMONG HEALTH CARE PROFESSIONALS (DOCTORS AND NURSES) WORKING IN EMERGENCY DEPARTMENTS IN MAJOR TEACHING HOSPITALS IN KANDY DISTRICT- SRI LANKA

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**Abstract**

**Background:** Pain is the most common reason for admission to the emergency department and effective pain management is detrimental for the outcome. The research focuses on assessing the knowledge and attitudes of pain management of healthcare professionals.

**Objectives:** The aim of the study was to assess the knowledge and attitude towards pain management among health care professionals working at major teaching hospital emergency departments in Kandy district.

**Methodology:** A cross sectional descriptive study was conducted at emergency departments of teaching hospitals in Kandy district and self-administered questionnaire were used for data collection.

**Results:** Total number of participants was 132 with 60.6% females. The majority (56%) were doctors which included 33.6% (44) medical officers, 12.2% (16) registrars and 6.1% (8) senior registrars in emergency medicine. Out of 44% nurses 18(13.7%) were trained in emergency and 40(30.5%) were not. Knowledge distribution of the sample has a mean of 12.61 with 2.449 SD out of 19 knowledge assessment questions. With a cut-off value of 50%, the majority (89%) had a good knowledge in pain management. Sample mean age was 40.08 ± 6.474 with 56% between 36-45 years of age. When considering clinical qualifications, MBBS qualified 39.4%, BSc in nursing 22.9%, Dip in nursing 20% with MD emergency medicine qualified 15.2% were there.

**Conclusions:** Position and the level of clinical qualification are significantly associated with knowledge and attitudes in pain management. The level of knowledge and attitudes on pain management is average. As the scarcity of the literature in Sri Lanka, further studies in the topic are recommended.

## Research Objectives

### General Objective

To assess the knowledge and attitude towards pain management among health care professionals working at major teaching hospital emergency departments in Kandy district -Sri Lanka

### Specific objectives

1. To describe the socio demographic details of healthcare care

professionals working at emergency departments in Kandy district

2. To describe the knowledge on pain management among health care professionals working at emergency departments in Kandy district
3. To describe the attitudes on pain management among health care professionals working at emergency departments in Kandy district
4. To identify potential barriers in pain assessment and management in emergency departments

## Methodology

A cross-sectional descriptive study was carried out at emergency departments of teaching hospitals in Kandy district and includes Kandy, Peradeniya, Sirimavo Bandaranaike special children hospital and Gampola hospital. All consented doctors and nurses, a total of 132 participants were recruited as study participants for the study. After obtaining the ethical clearance data collection was carried out using a self-administered questionnaire which was based on previous KAP studies on pain management and through extensive literature surveys [1-6]. The data were analysed through Statistical Package for the Social Sciences (SPSS).

## Results

The chapter discusses the sample's demographic features, attitudes and knowledge towards pain management and the factors that affect the knowledge and attitudes towards pain management.

The overall number of participants in the study was 132 with a mean age of 40.08 ± 6.474.

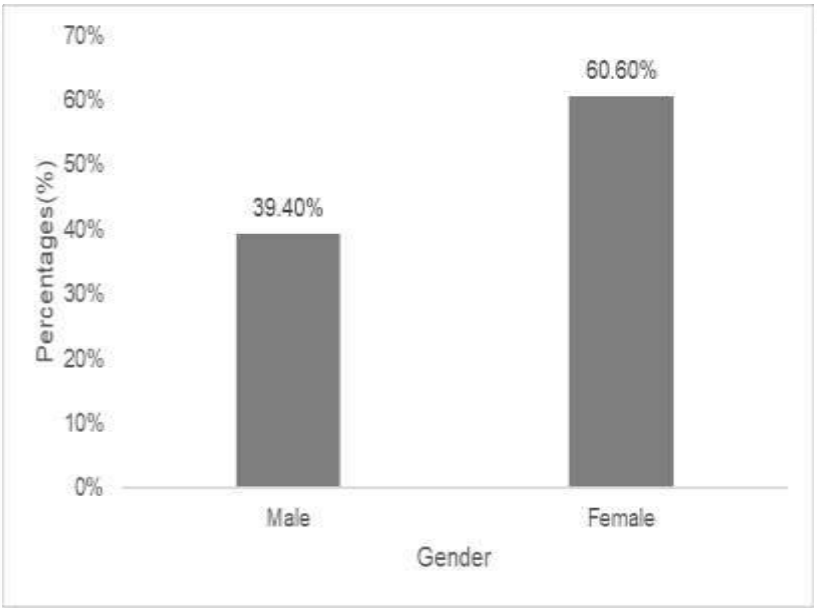


*Figure 1 - Gender distribution in the study sample*

The above column chart describes the gender distribution among the study sample. The sample was predominantly female (n=80, 60.60%) with males accounting for the remainder (n=52, 39.40%)

Table 01 - Distribution of highest clinical qualifications

| **Variable** | **Frequency (%)** |
|---|---|
| BSC in Nursing | 31(23.7) |
| DIP in Nursing | 27(20.6) |
| MBBS | 52(39.7) |
| MD Emergency Medicine | 20(15.3) |
| MRCP | 1(0.8) |

Different demographic factors and their frequencies obtained from the study population are described in the above table. Among the participants, the majority (52,39.7%) were MBBS qualified. 31 (23.7%) were having a BSC degree in nursing, 27 (20.6%) were having a diploma in nursing and 20 (15.3%) were qualified in MD emergency medicine.

Table 02 - Distribution of working position

| **Variable** | **Frequency (%)** |
|---|---|
| **Current employment within accident and emergency department** | |
| Acting Consultant | 2(1.5) |
| Consultant in emergency medicine | 2(1.5) |
| Consultant in general medicine | 1(0.8) |
| Medical officer | 44(33.6) |
| Nursing officer (Trained in Emergency) | 18(13.7) |

| | |
|---|---|
| Nursing officer (not trained in Emergency) | 40(30.5) |
| Registrar in emergency medicine | 16(12.2) |
| Senior registrar in emergency medicine | 8(6.1) |
| Years of experience in Accident and Emergency Medicine (Mean ± SD) | 5.88 ± 4.31 |
| **Continuous medical education (CME)** | |
| Yes | 69(52.3) |
| No | 63(47.7) |

Of the study sample, ⅓ (44) were working as medical officers while 40 (30.5%) nursing officers were working within the accident and emergency department with no additional training. 18 (13.7%) were emergency nursing officers and 16(12.2%) were registrar in emergency medicine. Mean years of experience in accident and emergency medicine of the study sample was 5.88 ± 4.31. Nearly half (52.3%) of the study sample (69) were engaged in continuous medical education.

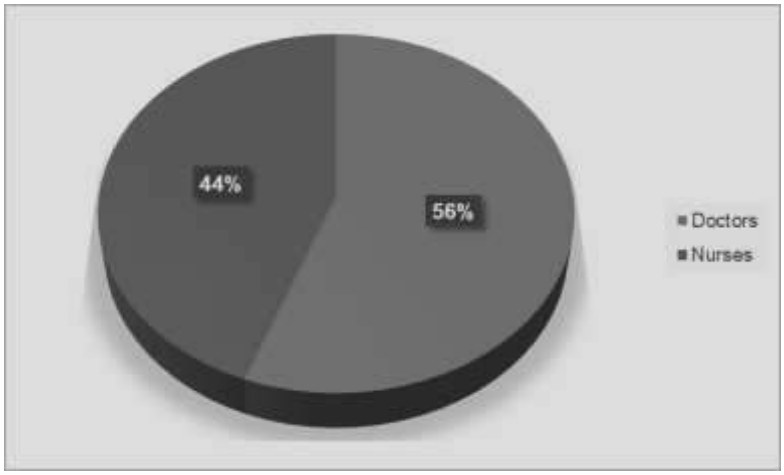


Figure 02: Distribution of Current position

Above pie chart describes the distribution of study sample according to the current position. More than half (n=73 ,56%) of the sample comprised of doctors with nurses accounting for the remainder.58(44%).

Table 03: Attitudes and practices assessment questions

| **Questions** | **True answer's frequency (%)** |
|---|---|
| <11 years old cannot reliably report pain, hence clinicians should rely on parent's pain assessment | 39(29.5) |
| <2 years of age have decreased pain sensitivity and limited memory of painful experiences | 39(29.5) |
| Until diagnosing the source of pain opioids should not be used | 56(42.4) |
| Staff can always pick up cues from children indicating pain | 69(52.3) |
| Vital signs are always reliable indicators of the intensity of a patient's pain | 43(32.6) |
| Narcotics should not be used in Pediatric patients as they can cause respiratory depression | 50(38.2) |
| Patients who can be distracted from pain usually do not have severe pain | 72(55.0) |
| Patients may sleep despite severe pain | 73(56.2) |
| Patients should be encouraged to endure as much pain as possible before using an opioid. | 53(40.5) |
| Patient's spiritual beliefs may lead them to think pain and suffering are necessary | 95(72.0) |
| Giving patients sterile water by injection (placebo) is a useful test to determine if the pain is real | 70(53.8) |
| A patient should experience discomfort prior to giving the next dose of pain meds | 60(45.5) |
| It is appropriate for the patients to request analgesics before the pain returns when they are receiving analgesics on a PRN basis | 98(74.8) |

| | |
|---|---|
| Children cry all the time; therefore, diversional activities are indicated rather than actual pain meds. | 69(52.3) |
| If you do not consider the condition to be painful the patient should not receive | 60(45.8) |

Above table describes the attitudes and practices towards the pain management. Of the participants 98 (74.8%) believed it is appropriate for the patients to request analgesics before the pain returns when they are receiving analgesics on a PRN basis. Most of the participants (n=95, 72%) had the opinion that a patient's spiritual beliefs may lead them to think pain and suffering are necessary. Nearly half of the sample (n=73, 56.2%) believed that patients may sleep despite severe pain.

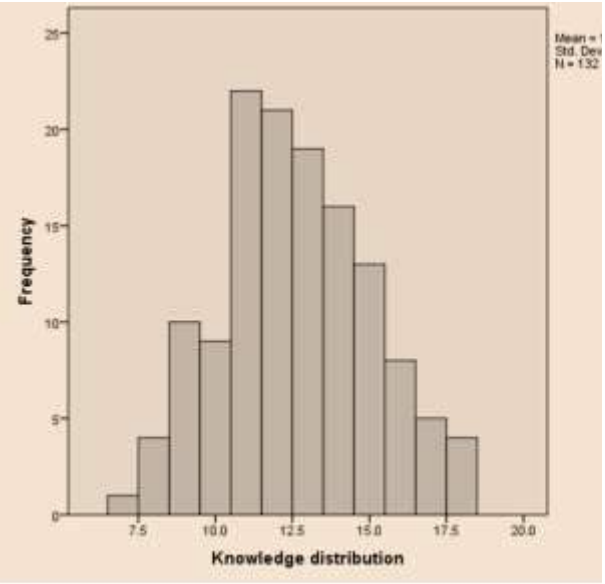


*Figure 3 - Knowledge distribution in the study sample*

This histogram illustrates the knowledge distribution in the study sample against the frequency. There were 19 true or false type questions assessing the knowledge. The knowledge distribution of the sample has a mean of 12.61 with a standard deviation of 2.449. The maximum knowledge value is 18, and the minimum is 7 in the study sample.

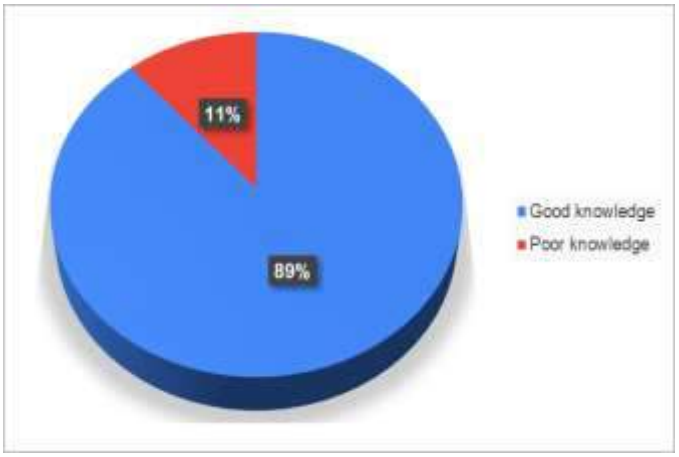


Figure 04: Distribution of knowledge (Good and Poor)

Above pie chart depicts the distribution of knowledge between 2 groups good and poor with a cut off value of 50%. The majority 117(89%) of the sample were having good knowledge and the rest 15(11%) were having poor knowledge.

Table 04 - Association between knowledge and demographic factors

| **Variable** | **Knowledge** | | **P Value** |
|---|---|---|---|
| | **Good (N=117)** | **Poor (N=15)** | |
| **Age** | | | |
| 25-35 years | 26(22.2) | 5(33.3) | - |
| 36-45 years | 67(57.3) | 7(46.7) | |
| >45 years | 24(20.5) | 3(20.0) | |
| **Gender** | | | |
| Male | 47(40.2) | 5(33.3) | 0.610 |
| Female | 70(59.8) | 10(66.7) | |

Of the 117 participants with good knowledge on pain management, 67(57.3%) were amongst the 36-45 age group. There were 15 participants with poor knowledge and of that 7(46.7%) were from the 36-45 age group.

Both groups with good knowledge and poor knowledge had the majority of female participation with 70 (59.8%) and 10 (66.7%) respectively. There is no statistically significant association between the gender and knowledge on pain management. (P=0.610)

Table 05: Association between knowledge and clinical qualifications

| **Highest clinical qualification** | **Knowledge** | | **P Value** |
|---|---|---|---|
| | **Good (N=117)** | **Poor (N=15)** | |
| BSC in Nursing | 25(21.6) | 6(40.0) | - |
| DIP in Nursing | 21(18.1) | 6(40.0) | |
| MBBS | 49(42.2) | 3(20.0) | |
| MD Emergency | 20(17.2) | 0(0.0) | |

| | | |
|---|---|---|
| medicine | | |
| MRCP | 1(0.9) | 0(0.0) |

Among the participants with good knowledge 49 (42.2%) were MBBS qualified whereas an equal number of participants with poor knowledge (6,40%) had BSC or DIP as their clinical education.

Table 06 - Association between knowledge and working position

| **Current employment within accident and emergency department** | **Knowledge** | | **P Value** |
|---|---|---|---|
| | **Good (N=117)** | **Poor (N=15)** | |
| Acting Consultant | 2(1.7) | 0(0.0) | - |
| Consultant in emergency medicine | 2(1.7) | 0(0.0) | |
| Consultant in general medicine | 1(0.9) | 0(0.0) | |
| Medical officer | 41(35.3) | 3(20.0) | |
| Nursing officer (Emergency) | 15(12.9) | 3(20.0) | |
| Nursing officer (not trained in Emergency) | 31(26.7) | 9(60.0) | |
| Registrar in emergency medicine | 16(13.8) | 0(0.0) | |
| Senior registrar in emergency medicine | 8(6.9) | 0(0.0) | |
| **Position** | | | |
| Doctors | 70(60.3) | 3(20.0) | 0.005 |
| Nurses | 46(39.7) | 12(80.0) | |

Of the group with good knowledge,41 (35.3%) were medical officers while 9(60%) of the group with poor knowledge were nursing officers with no emergency training.

The majority (70,60.3%) of the sample with good knowledge were doctors whereas 12(80% ) of the sample with poor knowledge were nurses. There is a statistically significant association between the position and the knowledge of pain management. (P=0.005).

Table 07 - Association between knowledge and Years of experience/ CME

| **Variable** | **Knowledge** | | **P Value** |
|---|---|---|---|
| | **Good (N=117)** | **Poor (N=15)** | |
| **Years of experience in Accident and Emergency Medicine** | | | |
| < 1 Year | 2(1.7) | 0(0.0) | - |
| 1-5 Year | 67(57.3) | 6(40.0) | |
| 6-10 Year | 37(31.6) | 6(40.0) | |
| >10 Year | 11(9.4) | 3(20.0) | |
| **Continuous medical education (CME)** | | | |
| Yes | 58(49.6) | 11(73.3) | - |
| No | 59(50.4) | 4(26.7) | |

Among those with good knowledge 67(57.3%) were having 1-5 years of experience in accident and emergency medicine. An equal percentage with poor knowledge (6,40%) had experience 1-5 years and 6-10 years respectively.

Nearly half of the participants with good knowledge 59 (50.4%) did not have continuous medical education (CME) and 11 (73.3%) of the participants with poor knowledge had CME. There was no statistically significant association with CME and knowledge of pain management. (P=0.103)

Table 08 - Association between knowledge and working hospital

| **Hospital** | **Knowledge** | | **P Value** |
|---|---|---|---|
| | **Good (N=117)** | **Poor (N=15)** | |
| Gampola hospital | 8(7.0) | 2(13.3) | - |
| Kandy general hospital | 56(49.1) | 8(53.3) | |
| Peradeniya teaching hospital | 34(29.8) | 4(26.7) | |
| Sirimawo bandaranayaka teaching hospital | 16(14.0) | 1(6.7) | |

Above table describes the association of knowledge and working hospital. Among those with good knowledge, 56(49.1%) were in Kandy hospital and 34 (29.8%) represented in Peradeniya teaching hospital.

Table 09 - Logistic regression

| **Variable** | **B** | **Sig** | **Exp(B)** | **CI 95%** | |
|---|---|---|---|---|---|
| | | | | **Lower** | **Upper** |
| Gender | -0.593 | 0.390 | 0.553 | 0.143 | 2.135 |
| Highest clinical qualification | -0.846 | 0.011 | 0.429 | 0.223 | 0.825 |
| Continuous medical education (CME) | -0.939 | 0.137 | 0.391 | 0.114 | 1.347 |
| Constant | 2.071 | 0.246 | 7.933 | | |

Logit p(x)=2.071- 0.593(Gender) - 0.846(Highest clinical qualification) - 0.939(CME)

Even if the independent variables (gender, clinical qualifications and continuously medical education) value is zero, the odds value can be computed as 2.071. When the gender increases by one unit, the logic p (x) is decreased by 0.593 units. Similarly, when clinical qualifications and continuously medical education rise by one unit, the logic p (x) value decreases by - 0.846 and - 939 respectively.

**Check the model significant; Hosmer and Lemeshow Test**

| Chi square | df | sig. |
|---|---|---|
| 4.193 | 8 | 0.839 |

H0: Model is adequate H1: Model is not adequate

Decision: If P value > 0.05, do not reject HO.

P > α ; 0.839 > 0.05

So do not reject HO

Conclusion: Model is adequate

So according to this model highest clinical qualification is the most significant variable for the knowledge.

## Discussion

Pain is an unpleasant sensory and emotional response resulting from actual or impending tissue damage [(7).] Pain, which is a global public health priority [(8),] has a higher prevalence both on admission and on the discharge from the emergency department [(9)] hence management of pain is paramount. For better management, knowledge of pain management among healthcare professionals is an essential component.

The sample was a female predominant sample with 60.6% of female participants out of total 132 with a mean age of 40± 6.474. The sample was collected from 4 hospitals in Kandy district. When considering the clinical qualification, the majority (39.7%) were MBBS qualified, 23.7% BSC in Nursing, 20.6% DIP in Nursing and 15.3% having qualified in MD emergency medicine. There were 33.6% medical officers, 30.5% nursing officers not trained in emergency,13.7% emergency trained nursing officers, 12.2% registrar in emergency medicine and 6.1% senior registrar in emergency medicine.

Similar studies have been conducted among emergency residents [(10),] physicians and nursing staff around the world [(11,12).] A study conducted in multiple hospitals in Jordan involved 60% nurses, 27% physicians, and 9% pharmacists [(11).] In contrast 56% of our sample was doctors and rest were accounted by nurses.

Using a questionnaire, the knowledge, attitudes, and practices towards pain management among the study sample were measured. Knowledge distribution has a mean of 12.61 with a 2.449 SD out of 19 knowledge assessment questions. With the cut off value of 50%, the majority (89%) of the sample had good knowledge. Most of the studies done were also using self-administered questionnaires [(13).] Also, there were studies done as quantitative experiments to assess the knowledge and attitudes towards pain management following educational sessions [(14).] The studies done among nursing officers

showed poor knowledge and attitudes towards pain management [13].

Among the participants with good knowledge the majority were medical officers (35.3%) while nursing officers not trained in emergency accounting for 26.7% and registrars in emergency medicine accounting for 13.8%. This can be due to shortage of trained staff to work in the Emergency Department (ED) and lack of training for the already working health care professionals. In the group with poor knowledge 60% were nursing officers with no training for emergencies and 20% each with emergency trained nursing officers and medical officers. Altogether of the group with good knowledge 60.3% were doctors and 80% of the group with poor knowledge were nursing officers which is similar to the literature where nursing officers having poor knowledge in pain management [11,12]. There was a statistically significant association between position and the knowledge of pain management (P=0.005).

Among those with good knowledge, 57.3% were having 1-5 years of experience in accident and emergency medicine. An equal percentage with poor knowledge (40%) had experience 1-5 years and 6-10 years respectively. As demonstrated before in literature specialized nurses have more knowledge scores but their years of experience did not seem to be related to that [15].

Of the study sample, 52.3% were engaged in CME. Nearly half (50.4%) with good knowledge did not have CME and 73.3% of the participants with poor knowledge had CME. A statistically significant association could not be seen between CME and knowledge of pain management. Even though this study does not show any association between CME and pain management, it is said to be improving physician performance and sometimes clinical outcomes [16].

Of the participants 74.8% believed it is appropriate for the patients to request analgesics before the pain returns when they are receiving analgesics on a PRN basis. Nearly an equal number of participants (72%) had the opinion that a patient's spiritual beliefs may lead them to think pain and suffering are necessary. More than half of the sample (56.2%) believed that patients may sleep despite severe pain. A study done in the United Arab Emirates shows nursing officers are having positive attitudes towards pain management [17]. Another study showed even though nurses had advanced appropriate attitudes towards management of pain, there are discrepancies between practices and attitudes. [18] Our study sample showed a mixed response between positive and negative attitudes towards pain management among the participants which is similar to the above findings.

Multivariate analysis showed a statistically significant association between highest clinical qualification and the knowledge of pain management. According to Tomaszek, better quality pain management is associated with continuous training and medical education [19]. Although this study was done among nurses and our study includes both doctors and nurses as participants, good clinical education is paramount for proper management in pain.

There are several barriers for identification and proper management of pain in the ED. Poor communication skills of patients and healthcare professionals, poor

interpretation of pain as it is subjective causes lack of identification of pain. Pain assessment has to be relied upon patient reporting as an objective measurement is lacking for measuring pain [20].

Similar to pain identification, managing pain has some barriers as well. Oligoanalgesia has been identified as a barrier for proper pain management in the ED [21]. Poor knowledge among healthcare professionals is also identified as another barrier [11]. We have identified that among our sample from ED there were nursing officers who did not train on emergencies, and they had poor knowledge about pain management.

## Conclusions

The level of knowledge and attitudes on pain management is average. Position and the level of clinical qualification are significantly associated with knowledge of pain management. Due to the scarcity of the literature in Sri Lanka, further studies on the topic are highly recommended.

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# A RETROSPECTIVE STUDY ON ADHERENCE OF DOCUMENTING CURB 65 IN OLDER PATIENTS WITH COMMUNITY-ACQUIRED PNEUMONIA AND PRESCRIBING ANTIBIOTICS ACCORDINGLY IN A TERTIARY CARE HOSPITAL IN THE UK.

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**Abstract**

**Background**: Community-acquired pneumonia is common in day-to-day clinical practice however can potentially be life-threatening. It is crucial for attending physicians to predict the prognosis and prescribe antibiotics according to available guidelines. CURB 65 is a tool widely used in clinical practice.

**Aim**: The study aimed to determine whether the clinicians adhere to documenting CURB 65 and prescribe antibiotics accordingly.

**Results**: Of 150 participants (females=80, 53.33%, The mean age was 87.1 years(4.9)), the CURB 65 score was documented in only 36(24.0%) patients' notes; however, antibiotics were prescribed according to the calculated CURB 65 score though it was not written in 61 patients (40.66%). Thus antibiotics were appropriately prescribed in 97 patients (64.67%); there is a statistically significant difference [Chi(1)=6.634, p=0.012] in the proper documentation and antibiotic prescription between the groups seen at the emergency department by the geriatric speciality team as a speciality vs the regular emergency team clinicians.

**Conclusions**: Documentation of the CURB 65 score when the diagnosis of CAP is made is highly inadequate. When the patient's were seen by a specialist team there was a statistically significant difference in documentation and prescription. However, antibiotics are prescribed according to local guidelines in nearly 65% of patients.

## Introduction

Community-acquired pneumonia(CAP) is a highly prevalent and potentially life-threatening infection, which has a poor prognosis in the elderly population and they are more prone to acquir CAP as well (1). The incidence of CAP 172.4 cases per 10 000 for adults aged 85 and over in the Netherlands(2) while 29.6 per 10 000 for all ages and 76.5 for adults aged 65 and over in Germany(3).

It is paramount important to treat them appropriately with proper antibiotics. CAP can be managed as an inpatient or outpatient depending on its severity. The CURB 65 score can determine the severity of community-acquired pneumonia and the national and local guidelines direct the treating clinician to prescribe antibiotics appropriately. Several predictors have been used to assess the prognosis of CAP. PSI pneumonia severity index(PSI) has

been used as the best tool while CURB-65 used as an alternative method(4).In patients admitted to hospital with CAP mortality is between 5.7 and 14%(5). Those admitted to an intensive care unit mortality are over 30% and more than half (60%) of pneumonia deaths occur in people older than 84 years(1).

CURB 65 is a clinical score, which includes the following parameters.

C - Confusion of new-onset (AMTS <8),

U - Blood Urea nitrogen more than 7mmol/l,

R - Respiratory rate more than 30 per minute,

B - Blood pressure less than 90 mmHg systolic or diastolic blood pressure 60 mmHg or less,

65- Age 65 years or older.

Depending on the CURB 65 score's magnitude, local guidelines (Norfolk and Norwich University Hospital) recommend appropriate oral or intravenous antibiotics. It is necessary to identify the severity by using CURB 65 score to determine the appropriate use of antibiotics and further management options(6). Consequently, that would prevent unnecessary use of antibiotics for the patient, which would further reduce the patient's morbidity and mortality.

Out of all the scoring systems currently being used in clinical medicine, the CURB-65 score is one of the most straightforward indices. Hence it is widely used in clinical guidelines for treating patients with CAP. Despite its simplicity, it is not being put into clinical practice. Findings of this study, which was conducted as a quality improvement project throughout the trust, would help to determine the depth of the problem of transferring academic guidelines into practice.

In Norfolk and Norwich University Hospital NHS Foundation Trust, old patients who are only over eighty years are consulted by Consultant Geriatricians. This age cut off has been adopted as per the demographics of Norfolk, which has a substantially higher percentage of a more ageing population, and nearly 10% of the population is eighty years and older(9). At given time geriatricians have to look after one-fifth of beds occupied by the older people of the hospital where 1200 beds capacity.

**Objectives**

1. To determine whether the clinician adhere to CURB 65 score documentation where community-acquired pneumonia diagnosis was made.
2. To determine whether the antibiotics were prescribed according to the CURB 65 classification using the patient's initial assessment and relevant laboratory investigations.
3. To determine where there is no documentation of CURB 65 score whether antibiotics were prescribed according to local guidelines.

**Methodology**

This study conducted using retrospective data from the emergency department of Norfolk and Norwich university hospital. Older patients were included in the study (above 80 years old). Data collection was done from May 2020 to December 2020. The study included data of all patients 80 years old and above who had been admitted to Norfolk and Norwich university hospital's emergency

department with a diagnosis of CAP from 01/02/1019 to 30/09/2019. The audit sample during the study included 150 patients. Patients were identified retrospectively by review of the coding system used in the emergency department.

Ethical approval was obtained by the Audit lead of the Norfolk and Norwich University Hospital and it was approved by the clinical governance team (Number-OPM 19-20-09)

Patients diagnosed with more than one diagnosis at the time of admission and who had other foci of infection were excluded. These patients were excluded as multiple diagnoses at the admission would directly affect the prescription of antibiotics and antibiotics choice. Similarly, other foci of infection were present simultaneously that directly influenced the selection and route of antibiotics administration.

Data collection was carried out from May 2020 to December 2020. The relevant data were obtained from electronic systems used in Norfolk and Norwich University Hospital.

Data analysis was conducted using SPSS version 25.0. Descriptive statistics using frequency distribution tables were used to present categorical variables and mean (SD) and median(IQR) values used to present continuous variables. Chi-squared test was used to assess the statistical significance of the subgroup analysis.

**Ethical considerations**

Approval was granted from the Norfolk and Norwich University Hospital audit monitoring committee to carry out the audit as planned. All necessary precautions were taken to store the data confidentially, and the audit was carried out in a strictly confidential manner by the principal investigator. A number identified each patient, and patient identifiable data was not used. Strict confidentiality was observed when the final report was submitted.

## Results

There were 150 patients in the study and the majority were females(n=80,53.33%). The mean age was 87.1 (4.9) years with a age range from 80 to 100 years, Figure - 01.

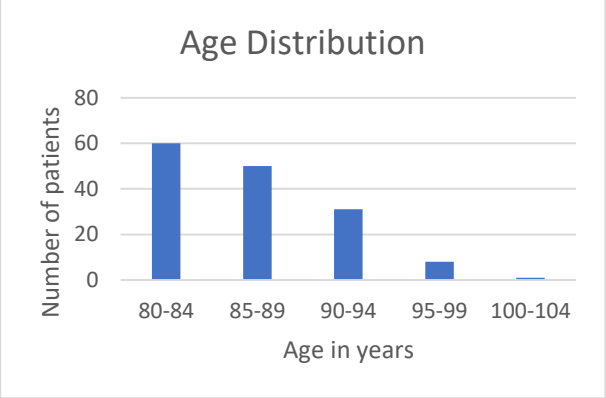


**Figure 1** – Illustrates the patients' age distribution and the majority (n=60,40%) were in the 80-84 age group.

CURB 65 scores were calculated in every patient in the audit irrespective of its documentation in individual patient notes. All scores were calculated at the time presentation to the emergency department by using electronic clerking sheets. CURB scores were from one to five, and the mean 2.5(SD) = 2.5(1.2). Median(IQR) was 2(2-3).

**Figure 2**- Show the number of patients who belong to each CURB 65 score from one to five. There were no patients who belong to score zero as the sample consisted of 80 years and above patients. There were 44 patients (29.33%) with CURB score two.

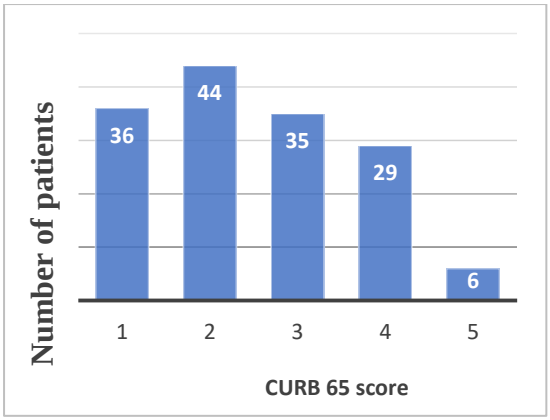


**Figure 2** -CURB 65 score characteristics

According to the Norfolk and Norwich University Hospital Trust guidelines, it was then audited whether the antibiotics were prescribed appropriately. Thus calculated CURB 65 score was used, and at the same time, it was noted whether CURB 65 was documented in the patient notes. Figure 3 illustrates documentation characteristics of CURB 65 score.

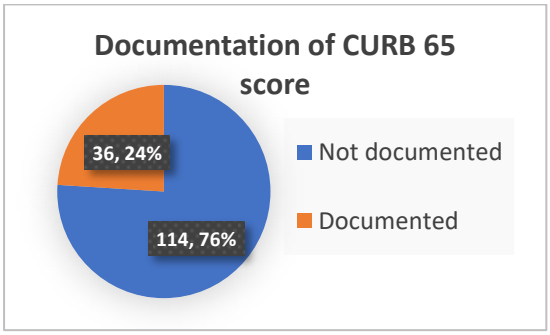


**Figure 3**- Documentation of CURB 65 score

Out of 150 patients, the CURB 65 score was documented in only 36(24.0%) patients' notes. However, antibiotics were prescribed according to the calculated CURB 65 score though it was not written in 61 patients (40.66%). Thus antibiotics were appropriately prescribed in 97 patients (64.67%).

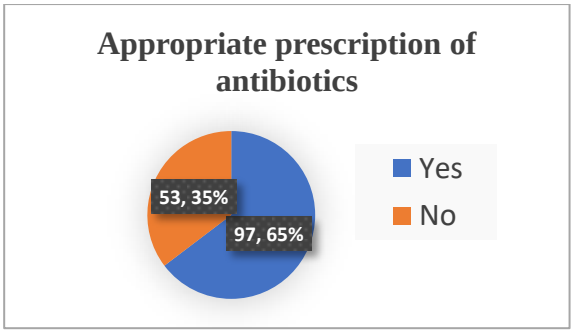


**Figure 4** Appropriate prescription of antibiotics

CURB 65 scores were documented, and appropriate antibiotics were prescribed, in other words, both requirements were full filled only in 36 patients(24.0%). Furthermore, all 36 patients in whom CURB 65 score was documented were prescribed appropriate antibiotics according to the local guidelines. (Table 1)

**Table 1** – Documentation and prescription

| Both criteria fulfilled | Number of patients | Percentage |
|---|---|---|
| Yes | 36 | 24% |
| No | 114 | 76% |
| Total | 150 | 100% |

Table 2 illustrates the number of patients in whom antibiotics were appropriately prescribed according to the calculated CURB 65 scores. However, there was no documentation of the CURB 65 score in the patient notes.

**Table 2**- Calculated CURB 65 score

| | Number of patients | Percentage |
|---|---|---|
| Prescribed but score not documented | 61 | 40.66% |
| Prescribed and Documented | 36 | 24% |
| None of the parameters met | 53 | 35.33% |
| | 150 | 100 |

Further, it was audited whether many of those patients in the sample were seen by Older People's Medicine team (OPM) as a speciality in the emergency medicine department itself (Table 3). Sixty patients (40.0%) were seen directly by older people's medicine team, and out of them, both documentation and prescription were fulfilled in twenty-one patients (Figure 5).

**Table 3-** Older people's medicine

| | Number of patients | Percentage % |
|---|---|---|
| Seen by OPM team | 60 | 40 |
| Seen by Main ED team | 90 | 60 |
| | 150 | 100 |

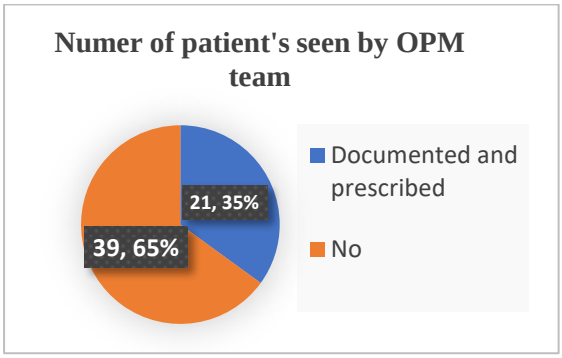


**Figure 5-** Documentation and Prescription of the patient's seen by OPM team

In addition, the number of patients seen by the night team was analysed and presented in table 4.

**Table 4**

| | Number of patients | Percentage |
|---|---|---|
| Day team | 75 | 50 |
| Night team | 75 | 50 |
| | 150 | 100 |

**Subgroup analysis**

CURB 65 score categories were divided into two categories for subgroup analysis. CURB 65 scores < 3 where the treatment guidelines recommend oral antibiotics. CURB 65 score > = 3 where treatment guidelines recommend intravenous antibiotics.

Subgroup analysis was performed on documentation and prescription of antibiotics in these two groups. The chi-squared test is used to determine whether there is a statistically significant difference between the groups.

**Table 5-Documented and prescribed Vs CURB 65 binary categories**

| **Prescribed and documented** | **CURB 65 score <3** | **CURB 65 score ≥3** | **Total** |
|---|---|---|---|
| **Yes** | **67** | **47** | **114** |
| **No** | **13** | **23** | **36** |
| **Total** | **80** | **70** | **150** |

There is a statistically significant difference [Chi(1)=5.645, p=0.022] in the proper documentation and antibiotic prescription between the CURB 65 score less than three group and the CURB 65 score of three or more group.

**Table 6- OPM team vs Regular emergency department team**

| **Documented and prescribed** | **Not seen by OPM team** | **Seen by OPM team** | **Total** |
|---|---|---|---|
| **No** | **75** | **39** | **114** |
| **Yes** | **15** | **21** | **36** |
| | **90** | **60** | **150** |

There is a statistically significant difference [Chi (1)=6.634, p=0.012] in the proper documentation and antibiotic prescription between the groups seen at the emergency department by the OPM team as a speciality vs the regular emergency team clinicians.

**Table 7- Day team vs Night team**

| Documented and prescribed | Day team | Night Team | Total |
|---|---|---|---|
| **No** | **53** | **61** | **114** |
| **yes** | **22** | **14** | **36** |
| | **75** | **75** | **150** |

There was no statistically significant difference [Chi(1)=2.339, p=0.180] in the proper documentation and antibiotic prescription between the groups seen.

**Discussion**

Given that proper documentation and proper antibiotic prescription according to local guidelines are highly recommended clinical practices, the audit was expected to have optimal clinical practice at the Norfolk and Norwich University Hospital.

Only 24% of the patients' CURB 65 score was documented though it was expected to be 100%. That was a common observation in where these sort of studies have done. A clinical audit conducted in the USA revealed that pneumonia bundle was only documented only in 47% of patients and even in the repeat audit, it was documented only in 53% of patients(10). It is imperative to document the CURB 65 score as antibiotic stewardship programmes depend on those data. CURB 65 score is vital to determine the treatment site, i.e., inpatient or outpatient and deciding what medications to be given as treatment(11). Hence, it is strongly recommended that clinicians document CURB 65 score once the diagnosis of CAP is made.

It was noteworthy to observe that despite not documenting the CURB 65 score, antibiotics were prescribed according to the guidelines in most instances according to the calculated CURB 65 score. Additionally, 61 patients (40.66%) had their antibiotics prescribed according to trust guidance though the CURB 65 score was not documented as expected. This made a total tally of antibiotics appropriately prescribed in 97 patients that means two thirds (64.66%) of patients had their antibiotics appropriately. Although, it was highlighted that documentation of severity scores would make communication between clinicians and effective identification of patients with early intervention and management clinicians do not document them as required(12).

Approximately one-third of patients, 53(35.33%) had no prescription of antibiotics appropriately. In these instances, it is harder to determine antibiotic treatment duration and why that decision was made by the initial clinician(13). That would further complicate continuity of care and existing antibiotic stewardship programmes (13). Evidence suggests that these practices would contribute to antibiotic resistance, additional economic burden, and complications like *Clostridium difficile* infection(14)(15).

The audit findings suggest that when it comes to CURB 65 documentation and

appropriate prescription of antibiotics if the patient was seen by older patients' medical team (OPM team) at the emergency department, the outcome was better. There were 60 patients (40%) seen by the OPM team, and out of those patients, 21 patients (35%) had both criteria fulfilled. Though the score was not documented, antibiotics were appropriately prescribed in 24 patients out of 60 patients seen by the OPM team. Hence 75% of patients had appropriate antibiotics been prescribed when the speciality team saw them. These data were compared with the patients seen by regular emergency department clinician, and there was statistical significance between two groups [Chi (1) =6.634, p=0.012]. This highlights that when the speciality team saw the patient, there was a better outcome. Any antibiotic optimization strategy depends on the clinician's experience and speciality (16). Efficacy and appropriateness of initial antibiotic therapy directly influence the patient's prognosis with CAP(17).

To assess how antibiotics choices are varied on the CURB 65 scores, CURB 65 categories were divided into two in which a score less than three and a score three or above. That division was made because intravenous antibiotics prescribed depend on whether the CURB 65 score equals or more than three and oral options are recommended when the score is less than three. There was a statistically significant difference in antibiotics prescription and documentation in these two categories ([Chi (1)=5.645, p=0.022]). This could be because of the initial use of intravenous antibiotics irrespective of the emergency department clinicians' CURB 65 score. There is a high propensity to use intravenous option initially irrespective of CAP's severity in the emergency department. This practice may adversely affect the patient's hospital stay, and sometimes patient who can be discharged may end up being admitted to the hospital. Of course, other factors are considered, such as unstable medical conditions, respiratory failure and social issues when deciding to bring the patient in as an admission(18).

In addition, a comparison was made during emergency department admissions during night hours and day hours; however, there was no statistical significance when it comes to documentation and prescription of antibiotics accordingly in those two groups. Due to institutional regulations implemented in response to COVID-19, there was an issue regarding the accessibility of patient records. Consequently, the audit sample hat to be limited to 150 patients.

## Conclusions

Documentation of CURB 65 score when the diagnosis of CAP made is highly inadequate. However, antibiotics are prescribed according to local guidelines in nearly 65% of patients, yet there is room for improvement.

When the patient is seen directly by the OPM team, there is a statistically significant difference in the CURB 65 documentation and appropriate prescription of antibiotics. Prescription is inappropriate when the CURB score is less than 3 when compared to CURB score is three or above, and this is statistically significant.

## Recommendations

A survey on medical staff to assess knowledge and practices of CURB 65

documentation and usage of local antibiotic guidelines for CAP is highly recommended.

Creating a drop-down option in the electronic documentation system to document CURB 65 score simultaneously, while noting the CAP diagnosis, would help facilitate the proper documentation practices.

As a part of antibiotic stewardship programme organizing a workshop on CAP and antibiotics for emergency department clinicians

A prospective audit to evaluate the improvement of documentation CURB 65 score and appropriate prescription of antibiotics following the dissemination of this audit findings is recommended.

In the Sri Lankan setting, it is vital to encourage juniors and seniors to adhere to documenting CURB 65 and prescribe antibiotics accordingly through continuous medical education.

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# A CASE OF SYNCHRONOUS DUAL METASTASIS TO COLON FROM BILATERAL ENDOMETROID TYPE OVARIAN CARCINOMA – AN UNREPORTED ENTITY

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## Abstract

Despite primary colorectal cancer being the third frequent malignancy worldwide, secondary metastasis to the large bowel from another primary cancer is a seldom reported. Only a handful of previous cases of primary ovarian cancer metastasizing to the large bowel have been described in literature published in English up to date. We report the management experience of a case of bilateral ovarian carcinoma with synchronous dual metastasis to the left colon, probably the first report of such nature. A fifty-six years old Sri Lankan female presented with non-specific left-sided abdominal pain for two months. She was found to have bilateral ovarian tumours with elevated serum CA 125 levels; splenic flexure and descending colon masses of adenocarcinoma on biopsy. The patient underwent total abdominal hysterectomy, bilateral salphingo–oophorectomy, left hemicolectomy with regional lymph node dissection and omentectomy. Histology confirmed endometroid adenocarcinoma of both ovaries with metastatic deposits in both colonic masses and in two out of seven lymph nodes along the left colic vascular pedicle. Both ovarian tumours and both colonic deposits were positively stained for ER and Vimentin; and were negative for CK20 on immunohistochemistry indicating ovarian origin. The patient is currently completing adjuvant chemotherapy at six months from the uncomplicated surgery. This case highlights the need for the surgeons to be mindful of similar rare occurrences of large bowel secondaries of ovarian origin especially when patients present with simultaneous large bowel and adnexal masses.

**Keywords:** Colorectal metastasis, Primary ovarian neoplasm

## Introduction

Colorectal cancer is the third frequent malignancy irrespective of gender, worldwide [1]. However, secondary metastasis to the large bowel from another primary malignancy is rare [1]. To the best of authors' knowledge, only seven previous cases of primary ovarian cancers metastasizing to large bowel have been reported in literature published in English up to date [1-8]. We report the management experience of a case of bilateral ovarian carcinoma with synchronous dual metastasis to the left colon, probably the first report of such nature.

## Case History

A 56-years old Sri Lankan female presented with left-sided abdominal pain for two-months. She never had altered bowel habits, bleeding/mucus passage per rectum or urinary symptoms. She was seven years post-menopause and had no gynaecological concerns. Over the preceding month, the patient developed loss of appetite, loss of weight, nausea, vomiting and symptoms of anaemia. On examination, the patient was pale and her body weight was 54kg (Body mass index-21.1kg/$m^2$). There was a non-discrete mass palpable in the left upper quadrant of mildly tender, non-peritonitic abdomen.

Her haematological and serum biochemical investigations were normal except for low haemoglobin level of 10.1g/dl. Ultrasonography of the abdomen showed a 12×9cm left hypochondrial solid mass close to splenic flexure. Further, there was a 6×4cm solid right adnexal mass and the left ovary was enlarged. Contrast Enhanced Computed Tomography (CECT) showed two large heterogeneous masses exophytically arising from splenic flexure (12×10cm) and distal descending colon (10×8cm). Further, Tomography confirmed bilateral adnexal masses (Right-6×4cm and Left-4×3cm) likely to be arising from ovaries (Fig. 1). There was no ascites, lung, liver, bony or nodal metastasis. Radiological evidences suggested bilateral malignant ovarian neoplasms and synchronous dual left colonic tumours. Lower gastrointestinal endoscopy detected a submucosal mass at descending colon, near completely occluding the bowel lumen which was biopsied. As the splenic flexure lesion could not be reached endoscopically, an ultrasonography guided percutaneous biopsy was taken. Both biopsies showed morphologically similar adenocarcinoma less likely to be of colonic origin. The patient had serum CA 125 levels of 3167U/ml [Normal 2-30U/ml] which raised the suspicion of a primary ovarian malignancy with rare metastasis to colon.

She underwent midline laparotomy under general anaesthesia. Operative findings were of two exophytic bowel masses, the larger at the splenic flexure measuring 12×11cm and the smaller at the descending colon measuring 11×8cm. There were bilateral ovarian masses which were apparently malignant comparable with tomography dimensions (Fig. 2A). There were no macroscopic secondaries in liver, omentum or peritoneum. The patient underwent total abdominal hysterectomy, bilateral salphingo–oophorectomy, left hemicolectomy with regional lymph node dissection and omentectomy (Fig. 2B). She made an uncomplicated recovery.

Histopathological examination of adnexal masses revealed endometroid type primary ovarian carcinoma in both ovaries. Uterus showed age related changes. Left hemicolectomy specimen showed two lesions at splenic flexure and descending colon with intact colonic mucosa (Fig. 2C). Microscopic findings were consistent with metastatic ovarian carcinoma favoring endometroid morphology containing vascular emboli in muscularis propria and serosa (Fig. 3). Overlying colonic mucosa and submucosa were devoid of tumour at both lesions and the omentum was free of tumour. Two out of seven lymph nodes along the left colic pedicle showed tumour infiltration. Both ovarian tumours and both colonic deposits were positively stained for ER and Vimentin; and was negative for CK20 on immunohistochemistry confirming the diagnosis of bilateral primary endometroid type ovarian carcinoma with dual colonic metastasis.

By the end of sixth month from surgery, the patient had successfully completed the six cycles of Platinum based combination chemotherapy and six out of the seventeen cycles of Bevasizumab.

**Discussion**

Despite primary colorectal cancer being the third frequent malignancy worldwide, secondary metastasis to large bowel from another primary cancer is seldom described. In such rare presentations, metastasis has been derived either from lung, ovary, breast, prostate, kidney or skin [1]. To the best of authors' knowledge, only seven previous cases of primary ovarian cancers metastasizing to the large bowel have been published in English as summarized in Table 1 [2-8]. The current case is unique, as this is the first description of bilateral ovarian carcinoma with synchronous dual metastasis to colon.

Transperitoneal seeding pathway has been postulated as the main mechanism for ovarian cancer metastasis to large bowel compared to hematogenous or lymphatic spread [1, 8]. That may explain the presence of exophytic masses in splenic flexure and descending colon in our case with tumour invasion being limited to the serosa and muscularis propria. All except one case of large bowel metastasis of ovarian cancer including the index case has been seen in the left colon and rectum [1,2,4-8]. Most of the previously reported cases have presented with non-specific symptoms and CECT and Magnetic Resonant Imaging have been used to assess the mass lesions in large bowel [1]. Endoscopic biopsies have been beneficial to obtain histology pre-operatively, especially in the cases with metachronous metastasis [2-4,6-8]. Immunohistochemistry has been the reference standard to differentiate colorectal secondaries from primary large bowel cancers. It is considered that CK20 and CEA positive stains to hint a carcinoma of primary colorectal origin and CK7, CA125, estrogen and progesterone receptor positive stains to indicate an ovarian origin, during the diagnosis process of large bowel metastasis of ovarian carcinoma [1].

This uncommon case highlights the need for the surgeons to be mindful of similar rare occurrences of large bowel secondaries of ovarian origin when a patient presents with simultaneous large bowel and adnexal masses.

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## Figures and Legends

Figure 1 – Coronal Computed Tomographic reformat of abdomen and pelvis (R- Right ovarian tumour, L-Left ovarian tumour, Outline arrow shows the mass in the splenic flexure and the solid arrow shows the mass in the descending colon)

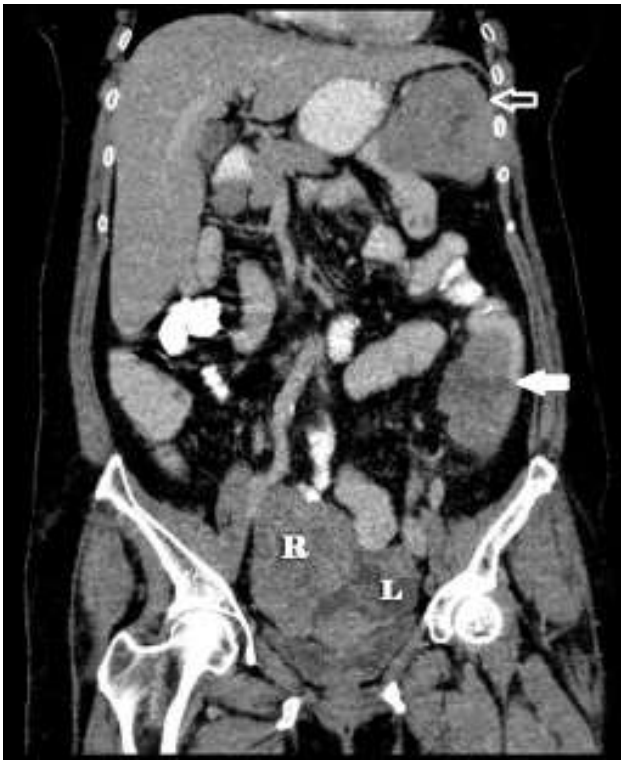


Figure 2 A – Perioperative image of the pelvis; B – Immediate postoperative specimens of left hemicolectomy with omentectomy and total abdominal hysterectomy with bilateral Salphingo-oophorectomy; C- Post Formalin fixated left hemicolectomy specimen cut opened to show the normal mucosa over both masses (Gray arrow – Ovarian tumours, White arrow – Lesion at splenic flexure, Black arrow – Lesion at descending colon)

Figure 3 – Photomicrograph of a colonic deposit to demonstrate tumour deposits in the serosa and muscularis propria preserving the mucosa (Haematoxylin and Eosin - ×4 Magnification)

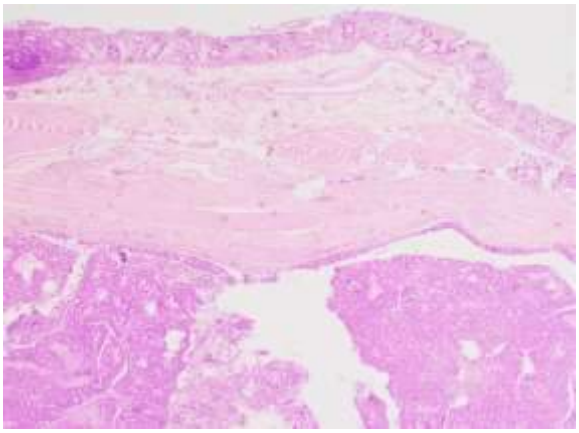

Table 1 - Summary of previously reported cases of ovarian carcinoma metastasizing to large bowel

| Author (Year) Country Reference | Age (Years) | Primary Pathology | Metastatic Site | Time gap for metastasis (Years) | Management Summary |
|---|---|---|---|---|---|
| Zighelboim et al. (2004) USA [2] | 39 | High-grade carcinoma with serous, endometroid, clear cell and focal mucinous components of Left ovary | Sigmoid colon | 0.5 | Initial left salphingo-oophorectomy; Subsequent right salphingo-oophorectomy, total abdominal hysterectomy, infra-colic omentectomy and sigmoid colectomy followed by chemotherapy |
| Shibahara et al. (2009) Japan [3] | 69 | Papillary serous type adenocarcinoma of Bilateral ovary | Caecum | 20 | Initial bilateral salphingo-oophorectomy, total abdominal hysterectomy, Ileac lymphadenectomy followed by chemotherapy; Subsequent right hemicolectomy |
| Yarze et al. (2009) USA [4] | 75 | Papillary serous type adenocarcinoma of Left ovary | Descending and Sigmoid colon | 6 | Initial bilateral salphingo-oophorectomy, pelvic debulking surgery, omentectomy followed by salvage chemotherapy and radiotherapy |
| Akhtar et al. (2012) India [5] | 50 | Papillary serous adenocarcinoma of Bilateral ovaries | Rectum | Synchronous | Bilateral salphingo-oophorectomy, total abdominal hysterectomy, ileac lymphadenectomy and left hemicolectomy followed by chemotherapy |
| Kim et al. (2015) Korea [6] | 70 | Papillary serous cystadenocarcinoma of Right ovary | Sigmoid colon | 8 | Initial bilateral salphingo-oophorectomy, total abdominal hysterectomy followed by chemotherapy; Subsequent anterior resection followed by chemotherapy |
| Park et al. (2018) Korea [7] | 74 | Serous adenocarcinoma of Left ovary | Sigmoid colon | 3 | Initial left salphingo-oophorectomy followed by right salphingo-oophorectomy, total abdominal hysterectomy, infra-colic omentectomy and chemotherapy; Subsequent anterior resection followed by chemotherapy |
| Aqsa et al. (2021) USA [8] | 53 | High-grade papillary serous carcinoma of Left ovary | Rectosigmoid junction | 2 | Initial bilateral salphingo-oophorectomy, total abdominal hysterectomy, infra-colic omentectomy and chemotherapy; Subsequent anterior resection followed by chemotherapy |
| Current study (2022) Sri Lanka | 56 | Endometroid type adenocarcinoma of Bilateral ovaries | Splenic flexure and Descending colon | Synchronous | Synchronous left hemicolectomy, bilateral salphingo-oophorectomy, total abdominal hysterectomy and Infracolic omentectomy followed by chemotherapy |

# A YOUNG GIRL WITH BILATERAL LOWER LIMB WEAKNESS

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## Abstract

Deficiency of vitamin B 12 is a metabolic disorder with number of causes. This often presents with megaloblastic anemia and neurological symptoms and signs which requires prompt treatment. But the diagnosis of vitamin B 12 deficiencies is challenging in resource poor setting and also due to rare occurrence in young people. A 22 year old lady who was a strict vegan, presented with progressively worsening bilateral feet and hand numbness with bilateral lower limb weakness for 5 months. Examination revealed pallor, glossitis and hyper pigmented skin mainly over knuckles, palmar creases, buccal mucosa and toes and graying of hair. Neurological examination revealed bilateral spastic paraplegia, exaggerated knee jerk and absent ankle jerk with equivocal plantar response. There was stocking type sensory loss in bilateral lower limbs up to mid-calf level with bilaterally impaired proprioception up to ankle joints. Clinical diagnosis of vitamin B12 deficiency was made with typical clinical features, hematological and biochemical parameters. Patient was treated with intramuscular and oral vitamin B12 and folic acid supplements. After one week of treatment patient's general condition was improved and after 6 weeks follow up, patient was able to walk alone with normalization of full blood count, retic count and blood picture.

High degree of suspicion of vitamin B12 deficiency in a patient presenting with peripheral neuropathic symptoms is important and blood picture evidence of hyper segmented neutrophils is a cost effective tool and a reliable way for diagnosis in resource poor setting.

**Key words**: Vitamin B 12 deficiency, Neurological examination, Blood picture

## Introduction

Vitamin B 12 (cobalamine) is a complex molecule in which a cobalt atom is contained in a corrin ring. Total body store of vitamin B 12 is 2 – 5 mg, of which half is stored in the liver. The recommended daily intake for adults is 2.6mcg/d and for children is 0.7mcg/d. As vitamin B 12 is highly conserved via the enterohepatic circulation, deficiency from malabsorption develops after 2 to 5 years. In vegetarians, deficiency from dietary inadequacy develops after 10 to 20 years [(1).]

Animal protein is rich in vitamin B 12. It's a water soluble vitamin and act as a coenzyme. It is important in cellular metabolism and maintenance of the integrity of nervous system [(2).] Vitamin B 12 is also important in DNA synthesis and thus in cell division [(3).] Therefore deficiency will lead to dyserythropoiesis and demyelination of nervous system [(4).]

## Case History

A 19 year old girl from farming community presented with insidious onset of pins and needle sensation in feet and hands which was progressively worsen over eight months period followed by lower limb weakness and difficulty in walking for recent five months. There were no bladder or bowel symptoms. She didn't have any other focal neurological

symptoms. She noticed darkening of skin over palmar crease, knuckles and toes. She had loss of appetite for last one month without any significant weight loss. Further, she noticed graying of hair and generalized hair loss since two years but no other clinical features of connective tissue diseases. She had no anemic symptoms or fever. She was a strict vegan since the age of 12 years.

On examination, her vitals were stable. Her BMI was 18.4 $kg/m^2$. She had pallor and glossitis. Neurological examination revealed bilateral spastic paraplegia with reduced power (1/5) in lower limbs with bilaterally exaggerated knee jerks and absent ankle jerks and equivocal plantar response. There was stocking type sensory loss to pain up to mid-calf level with impaired proprioception up to ankles. Upper limbs were normal neurologically with normal cerebellar signs and cranial nerves. Other systemic examination was unremarkable.

Blood tests showed hemoglobin: 10.4g/dl, Red blood cell (RBC) count: $3\times10^9$/L, reticulocyte counts: 0.4% and Mean Corpuscular Volume (MCV): 108fl. Platelet and white cell counts were normal. Blood picture showed macrocytosis and hyper segmented neutrophils which favoured vitamin B 12 deficiency without evidence of hemolysis or malignancy. Inflammatory markers were normal. Retroviral screening was negative. Serum electrolytes and creatine phosphokinase were normal. Liver and renal profiles were normal. Nerve conduction study showed no evidence of generalized large fiber neuropathy or Lumbosacral radiculopathy. Magnetic Resonance imaging of thoracic and lumbar spine was normal. Her vitamin B 12 level was very low (< 89pg/ml) (160 – 950pg/ml). Parietal cell antibody was not done due to lack of diagnostic tools. A clinical diagnosis of vitamin B 12 deficiency was made.

She was started on intramuscular vitamin B 12 1000u daily every other day for 2 weeks. Meanwhile physiotherapy started to minimize disuse atrophy. After 1 month of therapy her lower limb power was grade 4 and Hb count was 12g/dl with MCV of 95. Retic count was 3%. After 6 weeks of treatment she was able to walk alone and repeat serum vitamin B 12 was within normal limits. As she was responded to oral vitamin B 12 supplements, any absorptive errors of vitamin B 12 can be excluded retrospectively.

**Discussion**

Diagnosis of vitamin B 12 deficiency in resource limited settings is challenging as the access to diagnostic tool is limited and unfamiliarity of the disease especially among young population. Lack of intrinsic factor in patients with pernicious anemia is the most common aetiology worldwide and it is common in elderly. Other causes include malabsorption (gastric atrophy, achlorhydria, helicobacter pylori infection, intestinal bacterial overgrowth secondary to antibiotic treatment, long term ingestion of biguanides, antacids, $H_2$ receptor antagonists, and proton pump inhibitors; chronic alcoholism; gastric surgery/ bariatric surgery; pancreatic exocrine failure; sjogrens syndrome) and inadequate dietary intake [(1).]

Patients with vitamin B12 deficiency mainly present with neurological manifestations which include paresthesia, numbness, ataxia and limb weakness. Psychiatric manifestations also have identified which include irritability, decreased interest, delusions, sleep disturbances and depressive symptoms.

Diagnosis of vitamin B12 deficiency in a patient presenting with neurological symptoms is challenging as these neurological symptoms may be associated with several other diseases such as

diabetes mellitus, syphilis, HIV, alcoholism, other nutritional deficiencies like Copper, vitamin E and medications [5]. Laboratory investigations including full blood count and peripheral blood smear analysis are the mainstay of diagnosis which shows high MCV, pancytopenia, oval macrocytes and hyper segmented neutrophils. When compared to serum cyanocobalamine, blood smear evidence of hyper segmented neutrophils has a higher sensitivity in assessing vitamin B12 deficiency (sensitivity 90 – 95% and 98% respectively) [6, 7]. At present serum transcobalamine ii and methylmalonic acid levels are considered most specific indices in diagnosing vitamin B12 deficiency [8]. For diagnosis of pernicious anemia, anti-parietal cell antibodies and anti-intrinsic factor antibody are important.

Vitamin B12 replacement therapy can be done via either parenteral route or oral route. Having bound to intrinsic factor, vitamin B12 is absorbed actively in the distal ileum. However passive absorption 1- 2% of vitamin B12 also occur. Therefore, if administered at high doses, oral replacement can be effective as parenteral therapy [1].

MCV, RBC count and retic counts are important indices to assess the response and the therapeutic diagnosis. Therapeutic trials of vitamin B12 have also been used for the diagnosis as early diagnosis is essential for proper management and to prevent long term complications which can cause significant morbidity [9].

## Conclusion

High degree of suspicion of vitamin B12 deficiency in a patient presenting with peripheral neuropathic symptoms is important and blood picture evidence of hyper segmented neutrophils is a cost effective tool and a reliable way for diagnosis in resource poor setting.

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# SEVERE INVASIVE LIFE THREATENING COMMUNITY ASSOCIATED METHICILLIN RESISTANT STAPHYLOCOCCUS AUREUS INFECTION IN A NEONATE WITH PROBABLE PVL TOXIN PRODUCTION

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## Abstract

Community Associated Methicillin Resistant Staphylococcus aureus (CA-MRSA) are genetically distinct as they contain a novel cassette element, SCC mec IV and an exotoxin Panton Valentine Leukocidin (PVL). This toxin strongly associated epidemiologically with its virulence and explains pathogenicity. It predominantly causes recurrent skin and soft tissue infections (boils, abscesses, cellulitis), but occasionally causes invasive infections which can be life threatening (necrotizing pneumonia, necrotizing fasciitis, osteomyelitis, septic arthritis, pyomyositis) among previously healthy individuals in the community who lacks health care associated risk factors with no significant past medical history. We report a case of CA- MRSA in a15 days old baby who developed several severe invasive complications of PVL toxin production following CA-MRSA bacteremia such as bi lateral pneumonia, shoulder joint septic arthritis with associated right humerus osteomyelitis and deep intramuscular abscess, meningitis and large perineal abscess. Prompt diagnosis, adequate treatment and timely surgical interventions saved the life of the index baby in spite of suffering many serious sequelae.

**Key Words:** Methicillin Resistant Staphylococcus Aureus, Neonate, PVL Toxin

## Introduction

MRSA infection is caused by a type of staphylococcus bacteria that's resistant to many of the antibiotics used to treat ordinary staphylococcus infections. Two types of MRSA strains are noted, Health care associated (HA- MRSA) and CA-MRSA[1].CA- MRSA infections occurs among previously healthy individuals in the community who lacks health care associated risk factors with no significant past medical history whereas HA- MRSA typically seen in elderly, debilitated chronically or critically ill patients in health care settings. CA- MRSA are genetically distinct to HA-MRSA as they contain a novel cassette element, SCC mec IV and an exotoxin Panton Valentine Leukocidin (PVL). This toxin is strongly associated epidemiologically with its virulence and explains its pathogenicity which differs from HA- MRSA. It predominantly causes recurrent skin and soft tissue infections (boils, abscesses, cellulitis), but occasionally causes invasive infections which can be life threatening (necrotizing pneumonia, necrotizing fasciitis, osteomyelitis, septic arthritis and pyomyositis). CA- MRSA isolates are susceptible to many non β lactam antibiotics but in HA-MRSA choice of agents are limited due to resistance. Risk factors for transmission of CA MRSA infections are close contacts and crowded settings. Most cases are sporadic but, occasional clusters around close house hold contacts noted[2].

## Case History

A 15 days old baby transferred from a peripheral hospital to the special care baby unit of district general hospital with difficulty in breathing, fever, poor feeding of 1-day duration. Ante natal, post-natal histories were uneventful. On admission baby was febrile (103.1ºF), irritable and saturation on room air was 85%. Baby was connected to nasal prong oxygen, routine IV antibiotics for neonatal sepsis started and investigated further. On day 2, fever was continuing, swelling of right shoulder joint noted with warmth and was tender to touch. Baby was seen by orthopedic team and the diagnosis of right shoulder joint septic arthritis was made. X ray revealed an associated acute osteomyelitis of right humerus, and ultra sound scan picked up a deep intra muscular abscess formation. Platelets transfusion was done for progressive thrombocytopenia. On day 3, swelling over right perineal area was observed. Surgical team diagnosed it as a right perineal abscess involving labia majora. Blood culture taken on day 1 was positive for CA- MRSA. Same organism was isolated from day 3 blood culture and pus samples taken from joint aspiration and from perineal abscess. Antibiotics were changed immediately to 3 intravenous anti MRSA agents and stat dose of Intravenous immunoglobulin given. Prompt surgical and orthopedic interventions were done. On day 4, difficulty in breathing worsened and left segment cavitation with bi lateral consolidation noted in chest x ray. Baby was ventilated therefore. Baby was extubated after 4 days and was back on nasal prong oxygen. Same management was continued. When baby was clinically stable, lumbar puncture was performed. Cerebrospinal fluid full report was positive for meningitis and same antibiotics were continued as they cover meningitis. On day 26, left sided pneumothorax was seen on chest x ray and intercostal tube was inserted. After 35 days of admission baby was stable on room air. Baby was discharged after 41 days of hospital stay with anti MRSA oral agents. Clinic follow up was uneventful.

## Investigations

Blood investigations (on admission): WBC- 17 000, Platelets- 39 000, CRP- 219, ESR- 60.

Plain x ray- right shoulder joint- Acute osteomyelitis of metaphysis of humerus

Ultra sound – Right humerus osteomyelitis with deep intramuscular abscess

Chest x ray- Left - pneumothorax, cavitary segment, basal consolidation
Right- upper lobe consolidation

2D Echocardiogram, Ultra sound scan brain- No significant findings

Ultra Sound scan perineum- perineal abscess extending to perianal region

Blood/ Pus cultures - CA- MRSA isolated with sensitivity to ciprofloxacin, fusidic acid, teicoplanin, vancomycin, linezolid, cotrimoxazole and resistant to clindamycin, cloxacillin.

CSF full report: WBC – 27 cells (polymorphs- 44%), CSF culture- no growth

## Antibiotics

IV C. Penicillin / IV Gentamicin- 1 day

IV Cefotaxime- 6 days

IV Vancomycin/IV Clindamycin / IV linezolid – 21 days

IV Metronidazole – 10 days

O. Linezolid/ O. Clindamycin- 3 weeks (on discharge)

## Discussion

Only <2% of CA-MRSA and Staphylococcus aureus produce PVL toxin. These toxin producers rarely cause severe life threatening invasive infections in young healthy adults in the community, though it causes recurrent skin infections in majority[1]. It is not commonly encountered in neonates. Following CA- MRSA bacteremia, this baby has developed several severe invasive complications of PVL toxin producers such as bi-lateral pneumonia, septic arthritis of shoulder joint with right humerus osteomyelitis and deep intramuscular abscess, meningitis and large perineal abscess. Though the baby has all the clinical features suggestive of toxin production, we couldn't test for either toxin or gene due to lack of facilities in Sri Lanka.

Management of PVL producing CA-MRSA infection needs high clinical suspicion, early diagnosis, prompt initiation and continuation of 2-3 anti MRSA agents for longer duration to eradicate the infection completely, immediate transfusion of IV immunoglobulin to counteract toxin effect and prompt surgical interventions to drain focuses of infection[2]. Prompt diagnosis, adequate treatment and timely surgical interventions saved the life of this baby in spite of suffering many serious sequelae. Infection control aspect of management is also important in this disease[2]. Standard and contact precautions were strictly adhered, so none of the other babies acquired during stay. Contact tracing was done and none of the household contacts were known to be infected or colonized. So, source acquiring the infection to this baby was unknown. As the patient was a neonate it was decided not to do decolonization. Health education was given to the family to prevent re infection within the family.

## Conclusion

Early diagnosis, adequate treatment with antibiotics and timely interventions will improve the outcome of the infected patients. Targeted toxin studies would direct the proper management.

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# SPONTANEOUS CHOLECYSTOCUTANEOUS FISTULA: A RARE COMPLICATION OF GALLBLADDER DISEASE

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## Abstract

Spontaneous cholecystocutaneous fistula (CCF) is a rare complication of cholelithiasis, which has only been reported a few times in the literature. We report the case of a 59 years old woman who presented with a discharging sinus in the epigastrium. CCF is a type of external biliary fistula, which connects the gallbladder with the skin. Thilesus first described this phenomenon in 1670. There is usually a history of calculi in the gallbladder or neglected gallbladder disease. The incidence of CCF is rare, most patients being elderly females with the mean age of 72.8 years old.

Ultrasound, CT, MRI, MRCP and (CT or X-ray) fistulogram are used to confirm the diagnosis. CT was more significant than US in identifying the fistulous tract. Open cholecystectomy with excision of the fistulous tract is considered an acceptable option for treatment and it is curative in most cases. However, laparoscopic approach can be another option in experienced hands. Our patient underwent hepaticojejunostomy with en block skin, aponeurotic muscle, fistula tract, gallbladder, cystic duct, part of the common bile duct and hepatic duct excision.

This case report demonstrates that maintaining a high degree of suspicion of this rare entity is helpful in achieving correct preoperative diagnosis, and that computed tomography scan should be performed in all cases of unexplained abdominal wall suppuration or cellulitis.

**Key Words -** Cholecystitis, Cholecystocutaneous fistula, Hepaticojejunostomy, Fistulogram, Gallstones, Laparotomy, Bile ducts

## Introduction

Fistula is a pathological condition, which results from an abnormal connection between two epithelialized surfaces. Gallstone diseases predispose to biliary fistulas in rare occasions that connect between the biliary tract and other organs. Biliary fistulas are classified as: internal and external [1]. Internal biliary fistula connects the gallbladder with gastrointestinal tract and it results from chronic cholecystitis [2]. External biliary fistula connects the gallbladder with abdominal wall, it can be spontaneous, postoperative or post-traumatic or caused by iatrogenic injury of biliary tract [1,3].

Cholecystocutaneous fistula (CCF) is a type of external biliary fistula, which connects the gallbladder with the skin. In the medical history, it was first reported by Thilesus in 1670 and there are less than 100 cases of cholecystocutaneous fistulas have been reported. However, during that era, fistulas were a common complication of chronic and untreated cholecystitis [4]. Nowadays, the most rapid diagnosis and treatment with antibiotics or surgery significantly brought down the number of CCF cases. Apart from gallstone disease, CCF is reported in acalculous cholecystitis and gallbladder carcinoma.

The most common location for the exit tract of the fistula is the right upper

quadrant, but locations such as the gluteal region, umbilicus and right groin have also been reported [5]. CCF are mostly seen in women over 60 years of age but there are reported cases in younger patients as well. We report a case of a cholecystocutaneous fistula in a patient with previously undiagnosed gallstone disease.

**Case History**

A 59-year-old woman with the history of hypertension and dyslipidemia was admitted with complaints of pain in the abdomen and discharging sinus in the epigastrium for 2 years. She has had done an excision biopsy of epigastric sinus 1 year back at a different hospital and the histology revealed active chronic inflammation.

Ultrasound of the abdomen showed a 2cm tract in anterior abdominal wall in the epigastrium appears to communicate with the peritoneal cavity, in favor of fistula formation with background chronic cholecystitis and gallbladder calculi. Subsequently CT fistulogram was performed and findings were diagnostic of cholecystocutaneous fistula.

Liver profile was normal, and there were no contraindications for surgery under general anesthesia. We performed laparoscopy and decided to convert to open surgery due to difficulty in demonstrating the anatomy at the Calot's triangle. During laparotomy, the fistulous tract was demonstrated and found to enter the fundus of the gallbladder. Gallbladder was thick, fibrotic, and hard in consistency and was seen to adhere to the liver, with surrounding induration of the liver, and the cystic duct - common hepatic duct confluence, raising the suspicion of gallbladder carcinoma. There were several large gallstones within the gallbladder. We proceeded with en bloc excision of aponeurotic muscle, skin and fistulous tract together with the gallbladder, and a 5cm cuff of the liver, and cystic duct - common hepatic duct junction, and the common bile duct down to the level of the superior border of the duodenum. A jejunal Roux-en-Y loop was raised, and end to side hepaticojejunostomy performed with 5/0 Polydioxanone interrupted sutures. The drain was kept in subhepatic region and patient received broad-spectrum antibiotics during and after surgery. The patient expressed a slow but uncomplicated recovery and was discharged home well on post-op D6. Superficial wound infection was noted on D9, which was managed with oral antibiotics and by D20 wound was completely healed. Histology of the specimen revealed acute on chronic cholecystitis with a fistula between the skin and the lumen of the gall bladder, with no evidence of malignancy or tuberculosis.

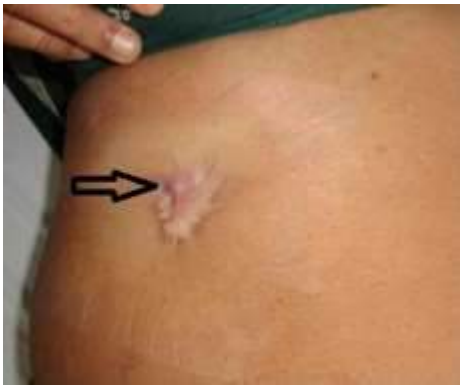

Figure 1 - Fistulous opening in the epigastrium

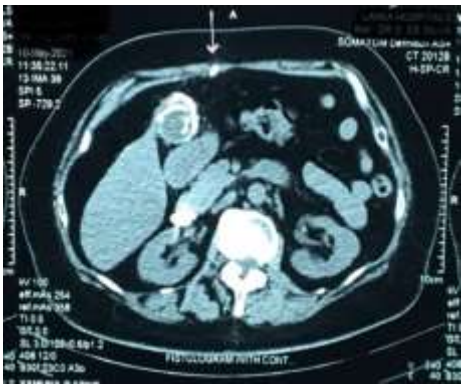

Figure 2 - Arrow showing the fistula

Figure 3 - 3D Constructed fistulogram

Figure 4 - Resected gallbladder with fistulous tract

## Discussion

The better understanding of pathology and evolution of sophisticated investigations lead to a rare occurrence of spontaneous cholecystocutaneous fistulas. Less than 20 cases of spontaneous CCFs have been reported over the past 50 years [5]. A neglected biliary tract disease is being the culprit of Cholecystocutaneous fistulas. They have been demonstrated at the umbilicus, left costal margin, right iliac fossa, right groin and the back [4]. The external opening of the fistula can mimic a pyogenic granuloma, infected epidermal inclusion cyst, chronic osteomyelitis of ribs, enterocutaneous fistula, discharging tuberculosis or metastatic carcinoma [4]. The nature of the discharge from the fistula can be purulent, mucoid or bile depending on the patency of the cystic duct.

Kaminsky reported on the frequency of biliary fistula to the gastrointestinal tract. The majority of the fistulas occur with connection to the duodenum (60%), followed by the colon (24%), stomach (6%) and choledochal duct (5%). Out of all, only 2% cholecystocutaneous abscesses or fistulas are accounted. Major risk factors for the development of spontaneous CCFs are: elderly females (>50 years), steroid therapy, history of typhoid, bacterial dissemination, trauma, immunocompromised conditions, etc.

The pathophysiology of CCF can be studied step by step in following manners: Cholecystocutaneous fistula is a sequalae of increased pressure in the gallbladder, secondary to cystic duct obstruction, either caused by a calculus or neoplasia. The intraluminal pressure slowly build up and leads to impairment of the blood flow and lymph supply to the gallbladder, thus causing mural necrosis and perforation. These fistulas, as presented in this case, most frequently arise from the fundus of the gallbladder. The state preceding spontaneous rupture has been termed “empyema necessitatis” by Nayman. This term essentially describes a “burrowing abscess” of the abdominal wall as a result of gallbladder inflammation.

Based on the underlying aetiology, the external biliary fistula management differs. The acute phase requires treatment with adequate antibiotics, analgesia and resuscitation. Not all external biliary fistula warrants surgical intervention because a proportion of patients exhibits spontaneous healing. So, in elderly or debilitated patients major interventions can be avoided. Possible surgical options include cholecystostomy with removal of the gallstones or cholecystectomy. Cholecystectomy is becoming the treatment of choice because cholecystostomy carries the risk of future stone formation in the gallbladder.

## Conclusion

Gallstones-disease related complications can be prevented by early laparoscopic cholecystectomy. In patients with anterior abdominal wall discharging sinus should warrant early referrals. Rare possibility of malignancy should be kept in mind while dealing with spontaneous CCF. In these cases, proper preoperative planning with imaging like CT scan is pivotal. At last, the choice of laparoscopic versus open and one-stage versus two-stage approach should be guided by the patient's clinical condition, local expertise and the best post-operative outcome for the patient.

## Consent

Written informed consent was obtained from the patient for the publication of the patient's clinical details and related images.

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# SUCCESSFUL PERIOPERATIVE ANAESTHETIC MANAGEMENT OF A PATIENT WITH DECOMPENSATED LIVER DISEASE FOR WHIPPLE'S PANCREATICODUODENECTOMY- INCORPORATION OF POINT OF CARE COAGULATION TESTING AT A RESOURCE LIMITED CENTRE

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## Abstract

Complications following major hepato-biliary resection surgery are high, even in specialized centres. Mortality and morbidity further increase in the presence of decompensated liver disease and coagulopathy play a significant role in such complains. Incorporation of point of care coagulation testing to manage perioperative coagulopathy during these surgeries is a novel phenomenon for optimal outcome. We present a case of successful perioperative anaesthesia with critical care management; and coagulopathy correction using point of care testing of a 77 year old male with a pancreatic head carcinoma and decompensated liver disease who underwent Whipple's pancreaticoduodenectomy at a resource limited centre.

**Keywords:** Point of care testing, Coagulopathy

## Background

Perioperative complications following major hepato-biliary surgeries in patients with decompensated liver disease are high and mortality rates of up to 50% have been reported [1]. To improve patient outcome, meticulous pre-operative assessment with risk stratification is essential and Child-Pugh score(C-PS) is used to prognosticate and predict the morbidity and mortality in cases with chronic liver cell disease. Whipple's Pancreaticoduodenectomy (WP) is a complex pancreato-biliary resection with many potential complications such as ileus, anastomotic leak, pancreatic fistula formation, coagulopathy with bleeding as well as thromboembolism [2]. Even though recent developments in surgical management have reduced mortality to less than 5% in centres of excellence, pancreatic resection is still accompanied by high morbidity rates (15%-20%) [2].

Pancreatic cancer is among the common malignancies associated with thrombosis affecting 50% of total patients [2]. Current recommendations strongly advise prophylaxis for venous thromboembolism in moderate to high risk patients awaiting major pancreatic surgery. However, there is a risk of bleeding during the perioperative period following WP increasing morbidity and mortality. The aetiology of such haemorrhage is multifactorial and treatment decisions are time sensitive. In the presence of the phenomenon of bleeding and thrombosis being present, point-of-care (POC) coagulation testing has a pivotal role in deciding perioperative anticoagulant therapy and transfusion of blood and blood products. Viscoelastic coagulation tests including thromboelastometry (ROTEM) and thromboelastography (TEG) have been recommended for assessment because the ROTEM/TEG-guided treatment algorithms reduced the transfusion of blood and blood products [3].

## Case History

Seventy-seven year old male with no significant past illness was scheduled for a WP with a diagnosis of T2N1M0 Pancreatic head adenocarcinoma after investigating for obstructive jaundice. He

was icteric and had a Body Mass Index of 21.1kg/$m^2$. Pre-operative assessment revealed deranged liver functions with CPS class B(Total Bilirubin – 649micromol/l, Direct Bilirubin – 307 micromol/l, Indirect Bilirubin -341 micromol/l, ALT – 208 IU/L, AST – 216 IU/L, ALP – 1135 IU/L, GGT – 556 IU/L, INR – 1.67, Serum Albumin - 3g/l). His Fasting Venous Glucose was 107mg/dl and cardiac assessment was satisfactory with a normal electrocardiogram and 2D Echocardiogram. The patients' coagulation profile showed a Bleeding Time-6 min, Activated Partial Thromboplastin Time (APTT) -31sec, Prothrombine Time(PT) – 19 sec, International Normalised Ratio-1.65. TEG revealed a hypercoagulable state (Figure 1). Even though TEG revealed a hypercoagulable state, only mechanical thromboprophylaxis was provided as the patient was fully mobile and risk of deep vein thrombosis was low.
Intra Operatively, Epidural line and invasive monitoring were established. Duration of surgery was 6 hours with estimated blood loss of 2500cc. In the absence of intra operative TEG monitoring, blood and blood products were administered on clinical assessment. Five units of Red Cell Concentrate (RCC), six units of Fresh frozen plasma (FFP) and six units of platelets were administered. Patient was haemodynamically stable throughout the intra operative period with an average urine output of 75 ml/hr.

Post operatively, the patient was electively ventilated for 24 hours and extubated on day 1. Oral clear fluids were started on same day. He was on Intravenous Cefuroxime 750mg and Metronidazole 500mg 8hrly. There was deranged coagulopathy during the post operative period (Table 1) as detected by conventional coagulation testing.

Table -1 Summary of conventional coagulation profile during postoperative period

| Date | PT(Sec) | INR | APTT(Sec) |
|---|---|---|---|
| Day 1 | 20.6 | 1.8 | 31.8 |
| Day 2 | 19.0 | 1.65 | 31.3 |
| Day 3 | 16.0 | 1.37 | 28.3 |
| Day 5 | 14.8 | 1.28 | |
| Day 6 | 23.02 | 2.05 | |
| Day 7 | 20.6 | 1.80 | 26.9 |
| Day 8 | 17.0 | 1.39 | |
| Day 10 | 15.2 | 1.29 | 28.1 |

Due to such derangement in coagulopathy attributed to decompensated liver disease, patient was transfused 2 units of RCC, 18 units of FFP, 4 units of Cryoprecipitate, 6 units of platelets in consultation with Transfusion Medicine after assessment of bleeding risk. Patient made a slow recovery over 12 post-operative days without any bleeding or thrombosis to be discharged on day 14.

**Discussion**

Significant functional reserve and nonspecific nature of liver blood tests affects the evaluation of the extent of liver dysfunction in patients with decompensated liver disease and hinders proper preoperative risk assessment. Conventional laboratory tests of blood coagulation only yield partial diagnostic information. POC devices are increasingly being used at the bedside for rapid, detailed testing of haemostatic function and for treatment monitoring in patients with coagulopathies[4].

Perioperative coagulopathies may necessitate the transfusion of allogeneic blood products and are an independent risk factor for perioperative mortality [2]. Blood clotting is conventionally tested with two global tests - INR, APTT, along with the platelet count and in some cases the fibrinogen concentration. This array of tests reflects only the initial formation of thrombin in plasma but not the details about clot stability or fibrinolysis. Furthermore, analysis at a standardized

temperature of 37° Celsius impedes the detection of coagulopathies induced by hypothermia. Test time of 40-60 minutes after blood drawing may not reflect the current state of the coagulation system and lead to inappropriate treatment[5]. All these limitations can be overcome by POC tests of coagulation. Despite not having institutional facility for TEG, we were able to perform preoperative TEG to assess the haemostasis. An intra operative TEG would have been beneficial to assess and minimize blood and blood products transfusion.

## Conclusion

Perioperative anaesthetic management of decompensated liver disease patient for major hepatobiliary surgery is challenging. POC coagulation testing can test various aspects of haemostasis rapidly in detail. Implementation of treatment algorithms based on POC coagulation testing may reduce the rate of transfusion of allogeneic blood products and reduce patient morbidity and mortality.

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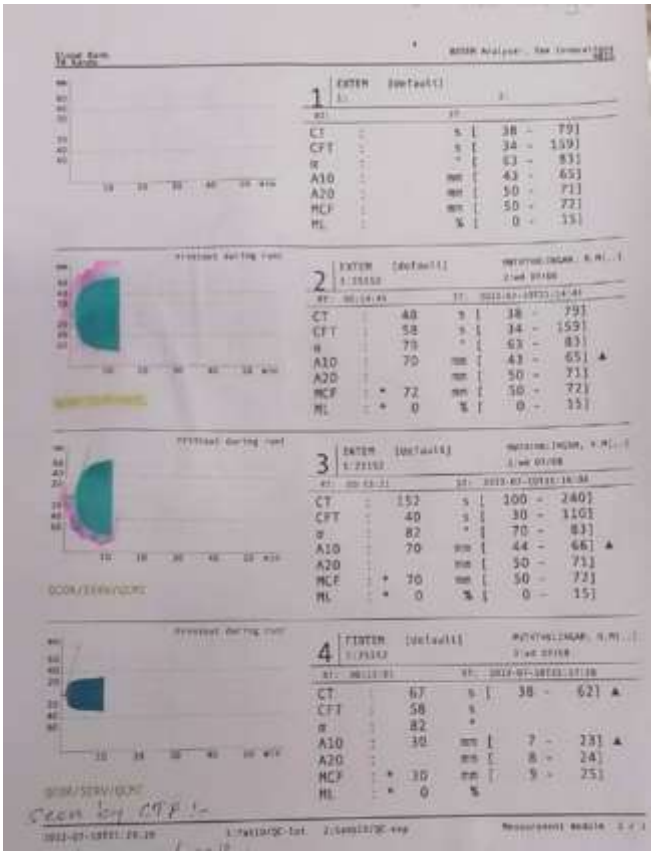
## Figure and Legend

Figure1:- Pre-operative Thromboelastogram



# A CLASSICAL PRESENTATION OF SCRUB TYPHUS IN THE NORTHERN PROVINCE OF SRI LANKA

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## Abstract

Rickettsial infection is an emerging disease which includes epidemic typhus, endemic (murine) typhus, scrub typhus, spotted fever group rickettsial infection and tick-borne lymphadenopathy (TIBOLA).The incidence is rising especially in the northern province of Sri Lanka. Clinical presentation can be variable and the presence of eschar highly suggestive of scrub typhus and will help to narrow the differential diagnosis. We describe a patient with classical presentation of scrub typhus who was treated with tetracycline with good clinical outcome. A high degree of suspicion is important for prompt diagnosis and thus to prevent complications of this deadly disease especially in resource poor setting where the serological diagnosis is limited.

**Key words**- Typhus, Eschar, Serology, Tetracycline

## Introduction

Rickettsial infection is an emerging disease of the tropics which was lethal in the pre-antibiotic era. This includes epidemic typhus, endemic (murine) typhus, scrub typhus, spotted fever group rickettsial infection and tick-borne lymphadenopathy (TIBOLA). Scrub typhus is caused by *Orientia tsutsugamushi*, an intracellular gram-negative bacteria that causes a significant burden in the Asia-pacific region and threatens one billion people [(1).] Transmission of scrub typhus requires the presence of trombiculid mites and rodents. Clinical presentation can be variable, ranging from mild illness to fulminant and potentially fatal disease. Clinical diagnosis is made with the typical presentation of fever, headache, maculopapular rash and the presence of eschar. There is an excellent response to tetracycline.

## Case History

A 38-year-old previously healthy male presented to the outpatient department with a 6-day history of fever, arthralgia, myalgia, headache and loss of appetite. The intermittent fever pattern is shown in figure 01. He has no other systemic symptoms and no high-risk sexual exposure.

He gave a history of camping out at night frequently during his long travels as a driver. On physical examination, the patient was febrile (39 °C). Abdominal examination revealed tender, soft splenomegaly of 3 cm below the left costal margin. There was non-tender, shotty lymphadenopathy in the cervical and inguinal region (figure 02).

Genital examination spotted an eschar over the right inguinal region. Laboratory tests revealed mild thrombocytopenia and mild transaminitis. His erythrocyte sedimentation rate was 56 mm/hr with a high C-reactive protein level of 45 mg/L. His chest X-ray, urine full report, cultures, retroviral and hepatitis screening were not significant. Malaria screening and 2D echocardiography were normal.

**Management and treatment outcome**

With a history of fever with arthralgia and myalgia, the patient was investigated for prevalent diseases in the island such as dengue fever, Covid-19 infection and leptospirosis. The finding of eschar and soft splenomegaly and the epidemiology data was consistent for scrub typhus. Unfortunately, serology testing was not available in the government sector laboratories at that time and the patient was started on a trial of doxycycline treatment. Prompt defervescence was observed after initiation of tetracycline which is also considered diagnostic of a rickettsial infection [(2).]

The patient had an unremarkable recovery and is under surveillance.

**Discussion**

Scrub typhus, also called bush typhus, is a mite-born disease caused by the bacteria, *Orientia tsutsugamushi*. Scrub typhus is spread to people through bites of infected chiggers (larval mites). This is a disease of the tropics mostly in South Asia, Indonesia, China, Japan and Nothern Australia [(3).]

There is a re-emergence of rickettsial infection worldwide and scrub typhus has an annual case prevalence of one million. The trend is seen in Sri Lanka as well [(4).] The number of notifications of rickettsioses in Sri Lanka showed an increment of 13-fold over a period of 20 years. Higher case detection may also contribute to this epidemiological rise of the disease. Data showed higher prevalence in the dry zone of the country. But the sero epidemiological mapping previously published did not include the northern region of the island, where the civil war and conflicts have underestimated the prevalence of the Northern Province [(5).]

Our patient had a classic presentation with an eschar. The finding of an eschar has a high diagnostic value clinically but it is not pathognomonic for scrub typhus where it is also found with ecthyma gangrenosum, cutaneous anthrax etc. The finding of eschar in females is mainly over the chest and abdomen whereas in males it is mostly found over the axilla, groin and genitalia. [(5,6)] Our patient had a history of fever, arthralgia, myalgia and splenomegaly where differential diagnosis such as dengue fever, infective endocarditis, malaria, typhoid and typhus were entertained. The finding of eschar was helpful in narrowing the differential diagnosis and initiation of treatment. The delay in diagnosis of typhus is detrimental where multi-organ dysfunction could occur involving myocarditis, pneumonitis, acute liver injury and acute kidney injury [(7).] The dramatic response to tetracycline was also diagnostic for rickettsial infection [(2).] Ideally, the

diagnosis must be confirmed with serology or repeated serology level of a four-fold rise in IgG levels. But unfortunately, serology testing was unavailable at that time in the government sector.

## Conclusion

Scrub typhus is an emerging disease in the world and also in Sri Lanka. More case prevalence should be expected in the northern province of the country than what the current data reveals. A high degree of suspicion is important for prompt diagnosis and thus to prevent complications of this deadly disease. The finding of an eschar is paramount in narrowing the differential diagnosis.

## Figures

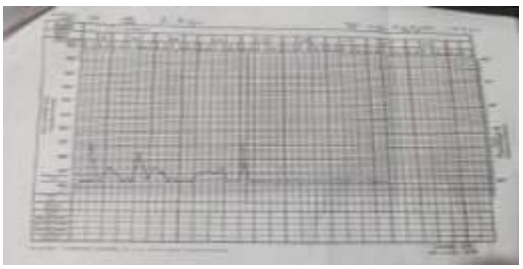

Figure1- Fever pattern

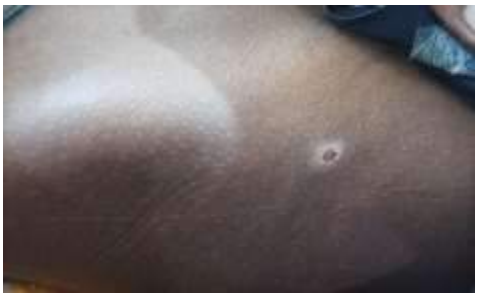

Figure 2- Eschar

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# MIDDLE-AGED FEMALE PRESENTING WITH FEATURES OF CHOLESTASIS

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## Abstract

Primary biliary cirrhosis (PBC) is a slowly progressive, autoimmune, cholestatic liver disease that should be differentiated from other causes of cholestasis by characteristic circulating anti-mitochondrial antibodies (AMA), histological findings, and biochemical features. A 52-year-old previously healthy female presented with clinical features of biliary obstruction. Biochemical investigations revealed marked elevations of aminotransferases and direct bilirubin, with moderate elevation of alkaline phosphatase. Infection screening was negative, and the extrahepatic biliary obstructions and space occupying lesions were excluded radiologically. The diagnosis of PBC was made by liver biopsy, positive anti-nuclear antibody (ANA) titer and AMA with elevated IgM levels. In middle-aged females with elevated ALP but no extrahepatic biliary obstruction, PBC should be suspected. AMA is the serologic hallmark of PBC and should be confirmed with a liver biopsy. It is associated with improved outcomes with early diagnosis and treatment. Follow up aims to detect complications like cirrhosis, hepatocellular carcinoma, and metabolic bone disease.

**Keywords:** Cholestatic liver disease, primary biliary cirrhosis, anti-mitochondrial antibodies

## Background

Primary biliary cirrhosis (PBC) is a slowly progressive autoimmune cholestatic liver disease that has characteristic features such as chronic cholestasis, circulating AMA, and histological findings of nonsuppurative destructive cholangitis with interlobular bile duct destruction. It should be suspected when a patient presents with chronic elevations of aminotransferases (AST, ALT), alkaline phosphatase (ALP), and total bilirubin with or without symptoms of primary biliary cirrhosis, such as pruritus or fatigue [(1)]. PBC is tenfold more common in women than men in their fifth or sixth decade of life [(2).] The etiology of this autoimmune disease was found to be related to genetic predisposition and microbial and xenobiotic environmental triggers [(3).] A considerable number of patients may progress to liver failure, liver transplantation, or death within ten years without identification and subsequent intervention [(1).]

**Case History**

A 52-year-old previously healthy female presented with severe loss of appetite with vomiting and abdominal pain associated with pruritus, fatigue and loss of weight for 2 months duration. On examination she was pale, icteric and had generalized hyperpigmentation and scratch marks with tender hepatomegaly.

Biochemical and immunological investigations were as follows.

| | | |
|---|---|---|
| ALP | 537.6 U/L | ( 98-258 ) |
| GGT | 173.5 U/L | ( 9 - 64) |
| ALT | 1074 U/L | (0.1 – 40) |
| AST | 1620 U/L | (0.1- 40) |
| Albumin | 30 g/L | (35 -50) |
| Total Protein | 77 g/L | (60 – 83) |
| Globulins | 47 g/L | (25 – 33) |
| Total Bilirubin | 367 µmol/L | (5 – 21) |
| Direct Bilirubin | 201 µmol/L | (0 – 3.4) |
| Indirect Bilirubin | 166 µmol/L | (3.4 – 12) |
| ESR | 52 mm/1$^{st}$ hr | (1- 20 ) |
| Hepatitis screening | Negative | |
| SAT | Negative | |
| ANA | 1:1000 | |
| Anti-mitochondrial antibody | Positive | |
| IgM | 185 mg/dL | (47 – 147) |

Abdominal USS revealed hepatomegaly with ascites and neither extrahepatic biliary obstruction nor space-occupied lesions. Probable causes for extrahepatic and intrahepatic cholestasis were excluded clinically, radiologically, and biochemically.

A liver biopsy confirmed the diagnosis of primary biliary cirrhosis, and she was treated symptomatically with ursodeoxycholic acid, vitamin K, spironolactone, etc. However, her clinical condition worsened over time. Later on, she developed features of cirrhosis. There was biochemical and hematological evidence of chronic liver cell failure, such as hypoalbuminemia, AST: ALT - > 2, elevated prothrombin time, and thrombocytopenia. She expired one year after diagnosing the disease due to severe hepatic dysfunction.

**Discussion and Conclusions**

Primary biliary cirrhosis is a rare, chronic, cholestatic liver disease with an autoimmune etiology. It can slowly progress into cholestasis with hepatic dysfunction and decompensation. However, the availability of more widespread immunological and biochemical tests has led to an increase in the diagnosis of PBC. PBC can be commonly associated with other autoimmune diseases and needs to be suspected in middle aged females who present with fatigue and itching. The classical features of abnormal liver function tests with typical cholestatic pattern of elevated ALP and the absence of extrahepatic biliary tract obstruction along with a positive AMA and liver biopsy can reveal the diagnosis.

PBC is associated with improved outcomes with early diagnosis and treatment with ursodeoxycholic acid. The only definitive treatment for PBC is liver transplantation. Follow-up aims to detect complications like

cirrhosis, hepatocellular carcinoma, and metabolic bone disease [2].

Factors associated with a poor prognosis include the presence of symptoms at the time of diagnosis, elevated concentrations of alkaline phosphatase and bilirubin, an advanced histological stage, the presence of antinuclear antibodies, cigarette smoking, and certain, genetic polymorphisms [4]. The risk of getting hepatocellular carcinoma is high in PBC with an advanced histological state, so all patients with primary biliary cirrhosis need to be screened for HCC using ultrasonography every six months, including testing for alpha-fetoprotein. [2]

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# MISLEADINGLY HIGH CA125 IN A PATIENT WITH TUBERCULOSIS

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## Abstract

Cancer Antigen (CA) 125 is a marker for epithelial ovarian tumours and endometrial carcinoma. CA125 can also be elevated in other causes of mesothelial cell activation such as inflammation. We report a 37-year-old lady, presented with nocturnal fever, sweats and shortness of breath for 2 weeks duration. Her computed tomography (CT) chest/abdomen revealed right sided pleural effusion with mild ascites. Diagnostic thoracentesis revealed blood stained, exudative pleural effusion with moderately high adenosine deaminase, marked mesothelial reaction and negative Ziehl neelson stain. Her CA 125 level was markedly elevated (1120U/mL). Biopsy of the right pleura was positive for acid-fast bacilli (AFB) polymerase chain reaction confirming the diagnosis of tuberculosis. After completion of anti-tuberculoid treatments, effusion was resolved and the CA125 levels were normalized. Markedly elevated CA 125 in this patient favours the diagnosis of malignancy in a background of negative markers for tuberculosis. Elevated CA125 levels are associated with tuberculosis, mainly in extra pulmonary locations.CA125 levels are useful in ruling in patients suspected of having tuberculosis with negative AFB. These uses of CA125 are yet to be studied in detail.

**Key words:** Pleural effusion, tuberculosis, CA125

## Introduction

CA125 is a glycoprotein found during the embryonic development of coelomic epithelium. It is a marker for ovarian and endometrial carcinoma.CA125 levels are elevated in > 80% of epithelial ovarian tumours. CA125 can also be elevated in other causes of mesothelial cell activation such as inflammation.[(1)]

## Case History

A 37-year-old, previously healthy lady, presented with nocturnal fever, night sweats and mild shortness of breath for 2 weeks. There was no history of loss of appetite or loss of weight. She is a health care worker.

Her general biochemical and haematological investigations were normal except moderately elevated CRP and ESR.

| Analyte | Result | Reference range | Unit |
|---|---|---|---|
| Full blood count | WBC :10.9<br>Neutrophils : 41%<br>Lymphocytes : **59%**<br>Haemoglobin : 11.4<br>Platelets : 230 | 4 – 11<br><br>11 -16<br><br>150 - 350 | $\times 10^3$ /µL<br><br>g/dL<br><br>$\times 10^3$ /µL |
| ESR | 45 | <20 | mm/hour |
| ALT | 13 | 10 – 40 | U/L |
| AST | 21 | 09 – 48 | U/L |

| | | | |
|---|---|---|---|
| Na+ | 138 | 135 – 145 | mmol/L |
| K+ | 4.5 | 3.5 - 5.3 | mmol/L |
| Creatinine | 63 | 40 – 60 | µmol/L |
| CRP | 28 | <6 | mg/L |

Chest x-ray (CXR) revealed a moderate right sided pleural effusion. High resolution CT (HRCT) scan of the chest and abdomen revealed mild right side pleural effusion and mild ascites.

Diagnostic thoracentesis revealed, blood stained pleural effusion with moderately high adenosine deaminase (ADA- 28U/L) and marked mesothelial reaction. However it was negative for AFB and malignant cells.

| **Test** | **Result** | **RI** | **Unit** |
|---|---|---|---|
| **Full report:** Appearance | Blood stained | Clear | |
| pH | 8.5 | 7.6 – 7.64 | |
| Cells: RBC<br>WBC<br><br>PMN<br>Lymphocytes | Numerous<br>6200<br><br>20%<br>80% | <10 | /cumm |
| Protein | 68.1 | 10 – 20 | g/L |
| Glucose | 81.8 | Similar as plasma | mg/dL |
| LDH | 580 | <50% of plasma | IU/L |
| | | | |
| Test | Result | RI | Unit |
| Cytology | No malignant cells, mesothelial cells>5% | | |
| ADA: | 113 | NL 15 ± 11<br>Pleural TB 86 ± 49 | U/L |
| Z.N.Stain | AFB not seen | | |
| AFB PCR and AFB culture | Negative | | |

Gold quantiferon test for tuberculosis was negative. Her circulating tumour markers showed markedly elevated CA 125 levels (1120U/mL) with normal CEA and CA 19.9.

Video assisted thoracoscopic wax biopsy was positive for AFB polymerase chain reaction (PCR) confirming the diagnosis of tuberculosis.

Thereafter she was treated with anti TB treatments with isoniazid, rifampicin, pyrazinamide and ethambutol for 6 months. After completion of therapy, patient's symptoms were completely resolved and the complete response of the pleural effusion was evidenced in CXR. The CA125 levels were normalized (37U/mL).

## Discussion

History of nocturnal fever, night sweats and mild shortness of breath together with right sided pleural effusion are suggestive of tuberculosis. Being a health care worker, contact history of tuberculosis cannot be excluded.

On investigation, moderately high ESR, blood stained exudative pleural effusion with lymphocytic predominance and high LDH together with high pleural fluid adenosine deaminase supports the differential diagnosis (DD) of tuberculosis (TB). However negative acid fast bacilli stain and culture and negative gold quantiferon test make TB unlikely as the main DD. In addition, marked mesothelial reaction in the pleural fluid is unlikely in a patient with tuberculosis.

Blood stained pleural effusion and markedly elevated CA125 of >1000 IU/mL together with mild ascites in HRCT abdomen raises the possibility of a malignancy rather than TB. Therefore, the patient was further investigated with whole body CT to exclude any evidence of malignancy elsewhere which was found to be negative.

Ultimately thoracoscopic biopsy was positive for TB PCR confirming the diagnosis of tuberculosis.

According to literature, elevated CA125 levels are associated with tuberculosis, mainly in extra pulmonary locations with abdominal involvement.[2] In this patient, both pleural effusion and ascites may have been contributed to the CA125 elevation. CA125 levels are useful in ruling in patients suspected of having tuberculosis with negative AFB. CA125 levels can also be used to differentiate tuberculosis from other pulmonary infections.[3] These uses of CA125 are yet to be studied in detail.

## Conclusion

Markedly elevated CA125 levels can occur in patients with tuberculosis especially in extra pulmonary locations with abdominal involvement. Usefulness of CA125 as a marker for tuberculosis should be further studied in detail.

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# RARE ADVERSE REACTIONS TO PLASMA COMPONENTS- ANAPHYLACTIC OR ANAPHYLACTOID?

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**Abstract**

Transfusion reactions are defined as adverse events associated with the transfusion of cellular component or non-cellular components (plasma, plasma derivatives). There is a growing concern that plasma utilization has increased over the years and many of them are inappropriate. Certain complications are more likely with plasma than other blood components. Non-infectious adverse transfusion reactions (NIATRs) like allergic transfusion reactions (ATRs) have emerged as the leading complication of transfusion. Anaphylactic/anaphylactoid transfusion reactions are rare but severe allergic reactions, associated with high morbidity and mortality if not detected and treated rapidly. To minimize hazardous out comes, obtaining proper history, thorough clinical evaluation and detailed documentation is of utmost importance.

We report a case of ?anaphylactic or anaphylactoid reaction to fresh frozen plasma (FFP) in a 74 years old male patient on warfarin , who was managed with FFP for warfarin induced upper gastrointestinal bleeding. During transfusion, the patient was noted to have a generalized urticarial rash with hypotension and hypoxia which responded well to intravenous medications with no adverse sequelae.

This review presents an overview of the clinical presentations of reaction, preventive measures and treatment modalities to successfully overcome an anaphylactic/anaphylactoid reaction.

**Key Words**: Adverse reactions, Anaphylaxis, Plasma products, Transfusion

## Introduction

ATRs are one of the commonest types of acute NIATRs usually occurring within 1-45 minutes from the start of transfusion, although the less severe allergic reactions may begin even up to 2-3 hours after the transfusion was initiated [1]. Ranging from mild (generalized urticaria, or localized angio-edema) to rapidly progressing severe life threatening anaphylactic reactions (with an incidence of 1:20000 – 1:50000 transfusions [2] which may lead to loss of consciousness, distributive shock, and in rare instances to death. Anaphylactoid reactions are clinically indistinguishable from anaphylactic reactions, but differ in their immune mechanisms. So a clinical definition, signs and symptoms alone cannot differentiate between these two [3].

## Case History

This 74 years old male on warfarin was admitted to medical ward due to hemoptysis and melena for 1 week. He is a

known patient with atrial fibrillation, ischaemic heart disease, heart failure and a tight aortic stenosis. He had no documented transfusion or allergic history. Following admission he was initiated on IV Cefuroxime.

Four packs of fresh frozen plasma (FFP) were requested and issued. During transfusion patient developed a generalized urticarial rash followed by hypotension ( BP -75/40mmHg ), hypoxia ( SpO2 – 92% ) with mild bradycardia (PR -60bpm) and was managed as anaphylaxis for FFP for which IM Adrenaline 0.5mg stat, IV Hydrocortisone 200mg and IV Chlorphenamine 10mg were administered. He was completely recovered following treatment. One and half packs were transfused over one hour period by the time reaction occurred. All the transfusion reaction serological investigations for red cells were normal.

Immunological investigations (IgA antibody, Anti IgA antibody, Serum Triptase) were sent to Medical Research Institute (MRI) for the confirmation of diagnosis. One pint of urgent blood transfusion was done (hemoglobin-7.1g/dl) with CPDA collected washed group specific compatible blood. Hypotension (BP -91/55mmHg) was noted 10 minutes after initiation. By that time the volume transfused was 75cc. Blood transfusion was immediately stopped and IM Adrenalin 0.5mg stat, IV Hydrocortisone 200mg and IV Chlorphenamine 10 mg were administered. Patient was stable with the management after reaction. Red cell serological investigations of post transfusion reaction samples were negative.

Second transfusion was done with group specific leuco depleted washed (more than 3 times) compatible blood before the time of expiry as hemoglobin was still 7.0 g/dl. Transfusion was initiated very slowly under strict observation after pre medicating with anti-histamines. No transfusion reaction was noted and blood transfusion was completed successfully.

**Discussion**

Anaphylactic or severe allergic transfusion reaction can be differentiated from other serious transfusion reactions by the presence of cutaneous symptoms, the absence of fever and chills, and the presence of severe respiratory symptoms [1]. Anaphylaxis characterized most significantly by hypotension and /or respiratory compromise typically occurring soon after initiation of transfusion.

Pathophysiology of acute immune mediated transfusion reactions is not well understood. The normal components of donor plasma may be immunologically different from those on the recipient.

Anaphylactic reactions are IgE – dependent responses (Type 1 immediate hypersensitivity reaction) of sensitized individuals to different allergens [3]. Anaphylactoid reactions are IgE independent responses, although the exact mechanism is unknown [1].

Plasma protein deficiencies play a major role in causing anaphylaxis. IgA deficiency (serum IgA <0.05mg/dl and normal IgG and IgM levels) is the commonest human immune deficiency [1]. Similarly individuals with haptoglobin deficiency, C4 or coagulant factor

deficiency, passive transfer of donor allergens (e.g peanut) or donor antibodies may also lead to anaphylaxis [4].

Some problems particularly associated with FFP transfusions are volume, citrate, temperature and sodium content.

To prevent future anaphylactic/ anaphylactoid reactions investigating on recipients' IgA levels and presence of anti-IgA antibodies are useful as these reactions can be life threatening in patients with IgA deficiency [5].

If an anaphylactic/anaphylactoid reaction is suspected, transfusion should be stopped immediately and supportive care should be provided including intramuscular epinephrine administration, IV fluid resuscitation, airway maintenance, and vasopressors if necessary. Steroids have a role in prevention of recurrence. Reaction has to be reported to transfusion services [4]. To prevent further reactions prophylactic pre-medication with anti-histamines is recommended [2]. If a patient with IgA deficiency with positive anti IgA antibodies needs blood transfusion, give RCC from unselected donors after double washing to reduce IgA levels below 0.005mg/dl [1]. For platelets requests give platelets from unselected donors after washing. For cryoprecipitate consider fibrinogen concentrates. Transfusion of the deficient plasma derivative (IV Immunoglobulin, Albumin, Rh immunoglobulin) is preferred over FFP. Coagulation factor correction should be done with pro-thrombin complex concentrates [1].

Regarding this patient presented here, he had no known history of allergy to food or drugs. Therefore cephalosporin which was administered earlier is unlikely to be the culprit. However since the post transfusion reaction red cell serological investigations revealed no evidence of a hemolytic transfusion reaction or serological in-compatibility, acute ATR to blood products was more likely with the presentation of the reaction. IgA levels sent to MRI detected within normal range. Even though the patient is not IgA deficient as the patient has experienced two anaphylactic reactions, transfusion of washed blood components may be preferred with pre medication, patient's consent and close medical supervision.

## Conclusion

Anaphylactic / anaphylactoid transfusion reactions are rare but severe life threatening non-infectious transfusion reactions and are successfully treated and easily preventable if identified on time.

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# CLOSURE OF RUPTURED SINUS OF VALSALVA ANEURYSM BY ATRIAL SEPTAL OCCLUDER DEVICE; A CHALLENGING CARDIAC INTERVENTION

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**Abstract**

A 61 years old male who had NYHA class II symptoms over one-year duration, presented with recent worsening of his symptoms. He had evidence of right heart failure with grade III continuous murmur along the left sternal edge. Trans thoracic 2 Dimensional echocardiogram followed by trans esophageal echocardiogram revealed a large ruptured aneurysm of non- coronary sinus of Valsalva into right atrium. While in the hospital stay, he was deteriorated with multi-organ dysfunction. Since he had progressive worsening of symptoms, a catheter based urgent intervention was carried out to close the defect. He had a remarkable symptomatic recovery immediately following the device closure and his Right Ventricular (RV) functions regained over the subsequent few months, achieving a normal RV and left ventricular function with no residual shunts at the end of one year follow up.

**Key words:** Ruptured sinus of valsalva, Atrial septal occluder device, Right heart failure

## Introduction

A Ruptured Sinus of Valsalva Aneurysm (RSVA) is an uncommon cardiac anomaly, either congenital or acquired in origin. Though Valsalva aneurysms can involve all 3 coronary sinuses, more often the non- coronary or right are involved in case of rupture, and that is more frequently happened to the right ventricle or right atrium[1].

This abnormality can be remained silently for many years and symptoms usually appear when the aneurysm ruptures into a cardiac chamber. That can induce a classical continuous murmur, exercise intolerance, symptoms of heart failure, or sudden cardiac death depending on the degree of the shunt[2]. Although the traditional treatment is surgical patch closure in this situation, we would like to share our experience in trans-catheter closure of RSVA in an emergency setting in a patient with severe Right Ventricular (RV) failure.

## Case History

A 61 years old male who had progressive exertional dyspnea over one-year duration presented with recent worsening of his symptoms. He had evidence of right heart failure with grade III continuous murmur along the left sternal edge. Trans thoracic

2 Dimensional echocardiogram showed marked RV volume overload, RV dysfunction and moderate pulmonary hypertension. More interestingly, there was a shunt from the aorta to right atrium. Trans Esophageal Echocardiogram (TEE) revealed a large ruptured aneurysm of non-coronary sinus of Valsalva into right atrium (*figure 01a and 01b*) with a continuous shunting of blood.

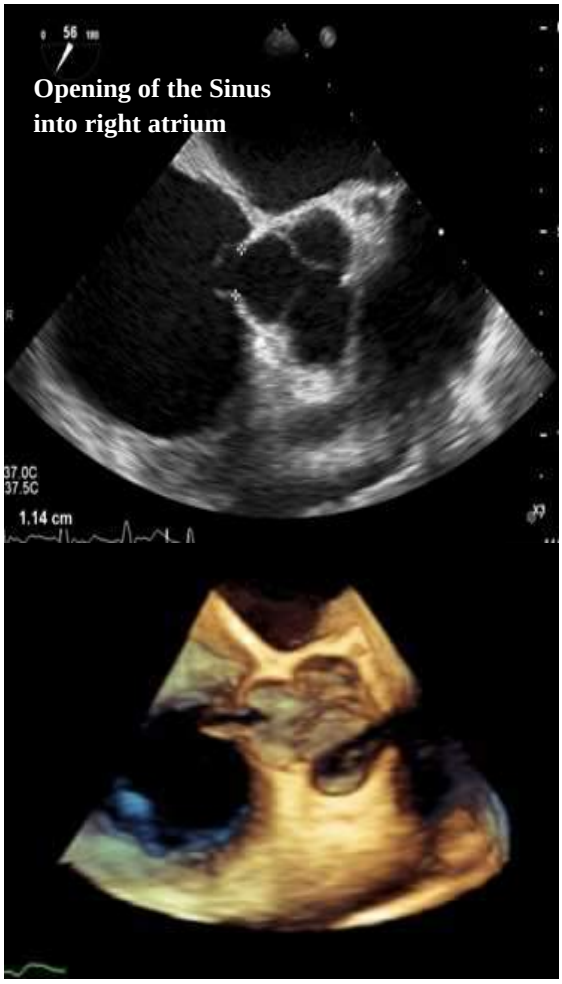



Figure 01a and 01b - TOE demonstrating large ruptured aneurysm of non-coronary sinus of Valsalva into right atrium

Subsequently, the aortogram confirmed the diagnosis of 9mm defect through the ruptured sinus *(figure 02)*. While in the hospital stay, he was deteriorated with congestive hepatopathy and development of cardio-renal syndrome. Since he had

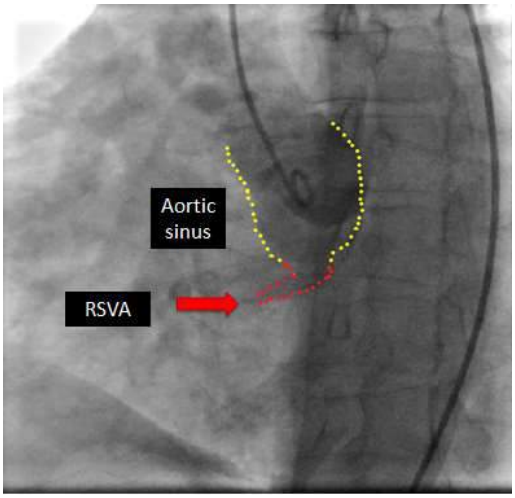



Figure 02 - Root aortogram before the device closure

progressive worsening of symptoms with multi organ failure, catheter based urgent intervention is decided over surgical closure by considering the high surgical mortality.

Antegrade approach was employed for device closure of our patient. Arterio-venous loop was created from the aortic side and the device with the delivery system was introduced from the venous side over the wire. The defect was closed with 13 mm Memopart® Atrial Septal Occluder (ASO) device (*figure 03*) since the available sizes of duct or Ventricular Septal Defect (VSD) occluder devices were not compatible with the size of the defect at that time.

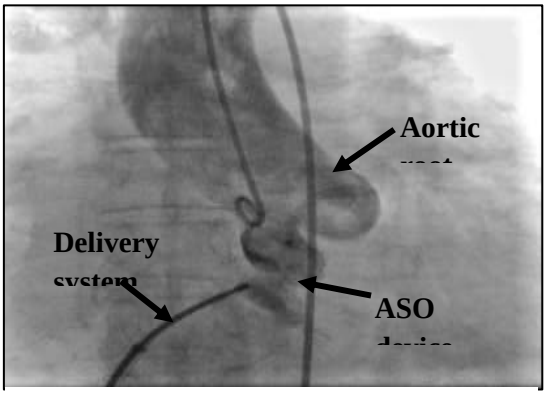
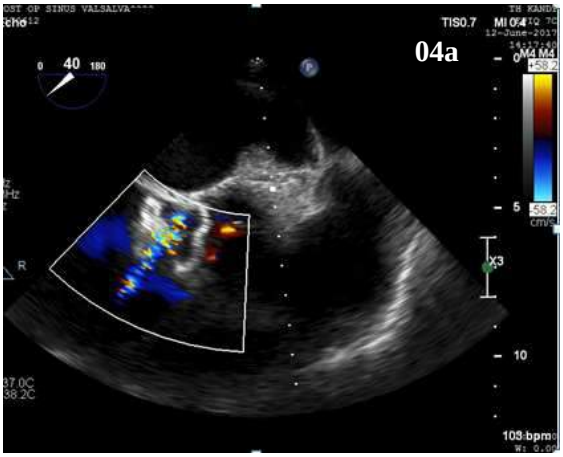



Figure 03 - Aortogram of the patient before the deployment of ASO device

The stability of the device was confirmed by fluoroscopy and trans-thoracic echocardiography while root aortogram confirmed the optimum position. After exclusion of impingement of the device on aortic leaflets, it was released under fluoroscopy guidance. Follow on TOE showed the device in proper position with minimal residual shunt (*figure 04a and 04b*).



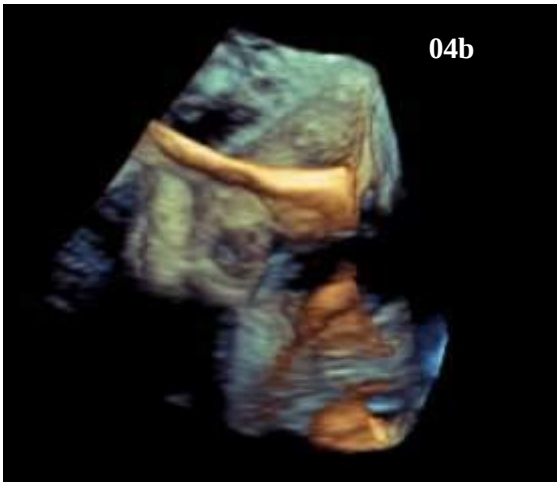




Figure 04a - Residual shunt at day one of the procedure
04b - 3D TOE demonstrating the optimal position of the ASO device

He had remarkable symptomatic recovery immediately following the device closure and his RV functions regained over the subsequent three months' period. After 12 months of follow up, he showed a normal RV and LV functions with no residual shunts.

**Discussion**

Sinus of Valsalva is an uncommon clinical entity with an incidence of 0.23% to 1.5% and interestingly, the rupture was observed among 35% of such cases[3]. Patients are usually asymptomatic until the rupture happens and some of the well-recognized aetiologies for such an occurrence are the strenuous effort, systemic hypertension, infection or trauma[1]. Other uncommon predisposing factors may be aortic valve surgeries, infective endarteritis, tuberculosis or even syphilitic aortitis[4].

The rupture of the sinus that results in acute left to right shunt will make the patient symptomatic as a result of acute RV volume and pulmonary arterial pressure overload. Once the deterioration of RV functions begins, the patient can be easily pushed towards the congestive hepatopathy followed by multi-organ dysfunction as happened in our patient. Therefore, the diagnosis of this condition should be made in correct time to avoid any potential complication that can occur in the subsequent disease course.

The diagnosis is generally established by trance thoracic 2D echocardiography with Doppler evaluation demonstrating the shunt position and direction. However, our experience on TOE suggests that it can be used as a minimally invasive, widely accessible rapid diagnostic tool even in a decompensated patient while Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) might further deteriorate the already deranged renal profile. TEE

can demonstrate the exact morphology of the RSVA, the size of the opening and the relationship of the defect to the aortic valve leaflets. Addition to that it also greatly helpful during the catheter based interventions for the device position and conformation of the device alignment. The pre-operative aortogram also yield much vital information that are helpful in the catheter based interventions. Here in our patient, we have planned our all the management decisions only depending on TOE and conventional aortogram results.

As the classical treatment approach, the open surgical closure of RSVA is considered as the treatment of choice[5]. However, as in our patient, the deteriorated RV function, congestive hepatopathy with high International normalized Ratio (INR), cardio-renal syndrome will make the surgical closure a risky procedure carrying high surgical mortality. Therefore, the device closure in this situation plays an utmost important role as a lifesaving intervention.

As the development of interventional cardiology, the initial approaches being Rash kind umbrella[6] and coils[7] to close the RSVA. However, the effectiveness and safety of ASO in catheter based interventions of RSVA was addressed in some of the previous studies that were conducted in recent past [3,8,9,10,11,12]. The most important advantage of such a procedure is that the minimal disturbance to the physiology that has already deranged in this type of decompensated patients. Here in our patient, the procedure was carried out under local anaesthesia without even sedation as he was in active liver failure. Other additional advantages of trans-catheter closure are the avoidance of medial sternotomy and Cardio Pulmonary Bypass (CPB), having shorter hospitalization and convalescence time.

There was a mild residual shunt noted through the ASO device initially following the device closure in our patient. However, with the time it was totally occluded and it may be as a result of the endothelialization of the device surfaces and it may facilitate the surrounding tissue stronger achieving a better long term sustainability of the procedure. Therefore, multi center studies with longer follow-up after such cases are needed to identify the correct prognosis of this type of interventions.

As with our experiences, trans catheter closure of RSVA is safe and effective especially in acutely decompensated patients with RV failure and even with multi organ dysfunctions where the open cardiac surgery using CPB carries high morbidity and mortality. However, a long-term follow-up is warranted to determine the enduring outcome of these patients following the catheter based interventions.

## Conclusion

Continuous left to right shunt from the RSVA may lead to progressive RV failure if not timely treated. Here we have demonstrated the possibility of RSVA closure by an ASO device in an emergency setting though the classical approach being the use of duct occluder or VSD closure devices.

## Consent

Informed written consent was obtained from the patient for the publication of this case report and any accompanying medical images.

**Conflict of interest** : The authors have no conflicts of interest to declare.

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# RITUXIMAB – THE SAVIOUR OF RESISTANT CUTANEOUS POLYARTERITIS NODOSA

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## Abstract

Cutaneous polyarteritis nodosa (cPAN) is a type of vasculitis affecting medium-sized vessels in the skin. In contrast, classic systemic PAN, cPAN is skin-limited.Most of the time, conventional immunosuppressive therapies are being used in the management, and very few case reports of refractory cPAN successfully treated with rituximab described in the literature.

**Key Words:** Rituximab, Polyarteritis nodosa,

## Introduction

Cutaneous polyarteritis nodosa (cPAN) is a type of vasculitis affecting medium-sized vessels in the skin, a distinct subset of polyarteritis nodosa (PAN). In contrast, classic systemic PAN commonly affects the kidneys, joints, muscles, nerves, and gastrointestinal tract, as well as the skin, vasculitis in cPAN is skin-limited[1]. We describe a patient with treatment-resistant cPAN successfully treated with rituximab.

## Case History

49 year old previously healthy shopkeeper presented with multiple painful crops of ulcers in both legs for 7 years. He denied any constitutional symptoms.

Examination revealed multiple punched out ulcers with a sloughy base over both legs (Fig 1). Peripheral pulses were normal. There were no clinical features of venous hypertension. Incisional skin biopsy from the edge of the ulcer showed necrotizing transmural vasculitis of small- and medium-sized vessels consistent with cPAN without evidence of granulomatous inflammation or fungal elements. ANCA, ANA, retroviral, cryoglobulin, Hepatitis B and C screenings were negative. Arterial and venous duplex scans of the lower limbs were normal.

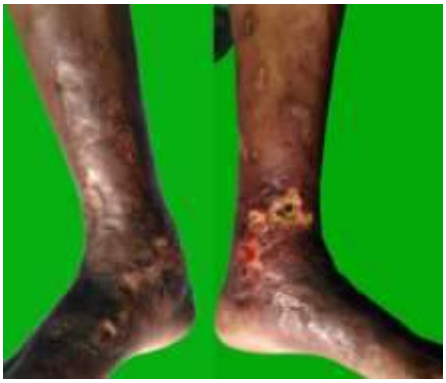

Fig 1 – Multiple punched out ulcers with a sloughy base

He was diagnosed with Cutaneous polyarteritis nodosa without systemic involvement.

Over the 5 years, he was on multiple immunosuppressants and oral prednisolone. Oral prednisolone was unable to tail off

below 20mg due to disease flare-up, which resulted in steroid-induced osteoporosis, hypertension, diabetes mellitus and obesity. Azathioprine, Methotrexate, Dapsone, Cyclosporine, Colchicine and Mycofenalate mofetil, were commenced as an adjuvant therapy along the course of illness but none was able to continue due to side effects of the drugs.

He responded well to Intra Venous Dexamethasone and cyclophosphamide pulses, and daily oral cyclophosphamide. However, it was withheld at the 4th pulse due to pyelonephritis and osteomyelitis. He also developed anaphylaxis to Immunoglobulin.

Local wound care was timely done. The ulcer did not respond to Topical potent corticosteroids or topical tacrolimus.

Rituximab was started after excluding contraindications. After the 1st cycle, he had a dramatic response and ulcers completely healed after 6 weeks (Fig 2).

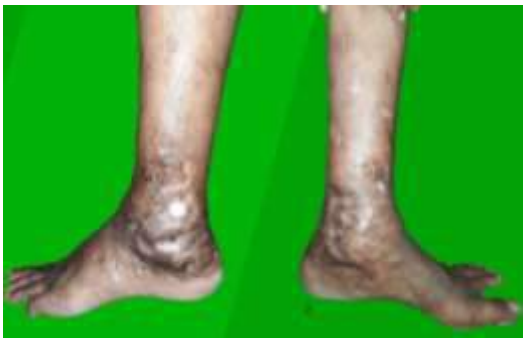

Fig 2 - Completely healed ulcers

## Discussion

Very few cases of refractory cPAN successfully treated with rituximab are described[2]. Data regarding the management of refractory cPAN are sparse in literature. Most of the time, conventional immunosuppressive therapies[3] are being used before biological agents like anti-TNF agents[4]. Our patient had refractory disease to all commonly used immunosuppressive therapies. Rituximab is a chimeric monoclonal antibody against the B cell-specific antigen CD20[5]; The treatment well-tolerated and effective in this case as ulcers completely healed within 6 weeks, he remains in remission up to now without, prednisolone or any other immune-suppressants for 12months. Thus, rituximab can be used for the treatment of refractory cutaneous polyarteritis nodosa.

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# LEUCOCYTOCLASTIC VASCULITIS AND PERIPHERAL ISCHEMIA AS FIRST MANIFESTATIONS OF SYSTEMIC LUPUS ERYTHEMATOSUS IN A YOUNG FEMALE

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## Abstract

Vasculitis is a known manifestation of Systemic Lupus Erythomatosus (SLE) even though it is not commonly occurring during the early course of the disease. Peripheral gangrene is considered to be very rare in SLE which can lead to severe ischemic necrosis and amputation. Peripheral gangrene usually occurs with a long-standing disease. Here we present a 26 years old previously healthy female presenting with erythematous rash in legs with edema and necrotic skin lesions over nostril and left ear pinna with ischemic toe tips. Her serology was positive for SLE and diagnosis of lupus vasculitis with peripheral ischemia was made. She markedly responded to steroids and immunosuppressant. Vasculitis with peripheral ischemia as a first presentation of SLE is rarely found in the literature. Early diagnosis and treatment is necessary to prevent tissue loss.

**Key Words** Systemic Lupus Erythematosus, Vasculitis

## Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease of unknown etiology. It can affect any organ system in the body by producing autoantibodies which induce inflammation (1). SLE has various presentations depending on the organ system involved but usual presentation includes malar rash, arthritis, and fever.

Vasculitis is a well-known manifestation of SLE. It can present with various clinical forms depending on the size of vessel involved. Estimated prevalence of vasculitis in SLE is range from 11 to 36%. Small vessel involvement is the most common, but medium and large vessel involvement also has been identified (2).

Peripheral ischemia and gangrene is considered to be very rare in SLE which is 1 to 3% of SLE patients. This can lead to severe ischemia, necrosis and amputation.

## Case History

A 26 years old female who was previously healthy presented with 4 days history of painful erythematous rash involving legs with gradual involvement of hands and forearms. She had blackish discoloration of nostrils (figure I) and left ear lobe (figure II) as well. Meanwhile she noticed leg swelling and painful bluish discoloration of toe tips (figure III). She denied features of Raynaud's phenomenon. She had experienced constitutional symptoms for 2 weeks prior to the appearance of rash. She had no other

clinical manifestations of SLE or connective tissue disease in the past or presently. On examination, she had palpable purpura with evidence of ischaemia in bilateral toe tips. Her hemodynamic was stable. Other systemic examination was insignificant. Her immediate blood work up showed very high Erythrocyte Sedimentation Rate (ESR) (123mm/1$^{st}$ hour) and C- reactive protein level (119mg/dl) with neutrophil leukocytosis (white blood cell 13x10$^{3}$/µl, neutrophil 10x10$^{3}$/µl, lymphocyte 1.8x10$^{3}$/µl), mild thrombocytopenia (110x10$^{3}$/µl) and anemia (8.7g/dl). Blood picture showed inflammation without evidence of hemolysis. Coombs test was negative. Septic screening was negative and procalcitonin was negative. Hepatitis screening and retroviral screening were negative.

Urinalysis showed no active sediments. Her other systemic assessment revealed no significant abnormality including chest radiography, ultrasound abdomen, 2D echocardiography and renal functions. Bilateral lower limb venous duplex was normal and arterial duplex showed no obstruction.

Antinuclear antibody was significantly positive (1/1280). Anti dsDNA antibody was negative. She had low complements 4 (C4) (6mg/dl) (12 – 36mg/dl) and low normal complement 3 (C3) levels (85 mg/dl) (83 – 177mg/dl). Anticardiolipin antibody IgG was weakly positive (13.18 U/ml) and IgM was positive (19.95 U/ml). Rheumatoid factor was negative. Skin biopsy showed evidence of leukocytoclastic vasculitis with vessel injury and some vessels containing fibrinoid material.

With above manifestations and serological markers, she was clinically diagnosed with SLE with lupus vasculitis (leucocytoclastic vasculitis) and impending peripheral ischemia. She was started on 1000mg of methylprednisolone pulse therapy for 3days followed by oral prednisolone 1mg/kg/day. Meanwhile she was started on hydroxychloroquine 200mg/day and anticoagulation as well. Currently she is on tailing-off dose of Prednisolone, Methotrexate and low dose Aspirin.

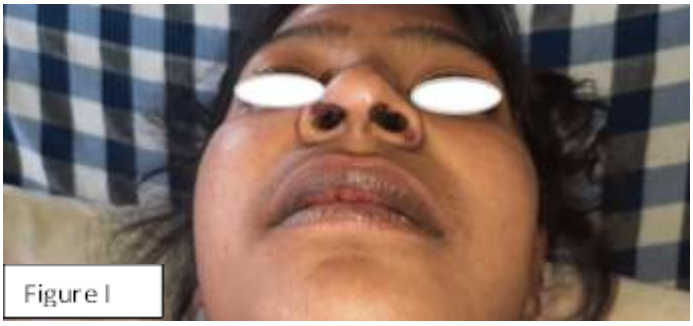

Figure I

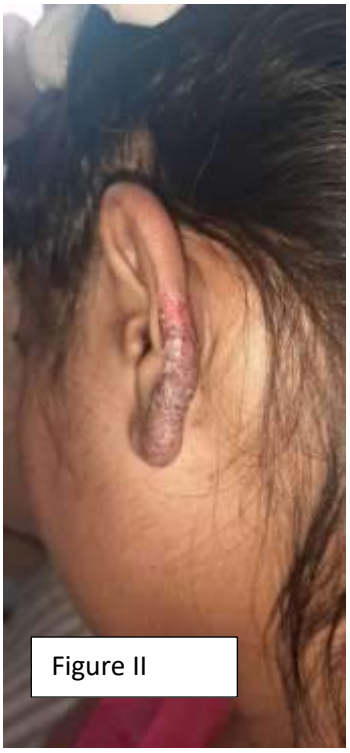

Figure II

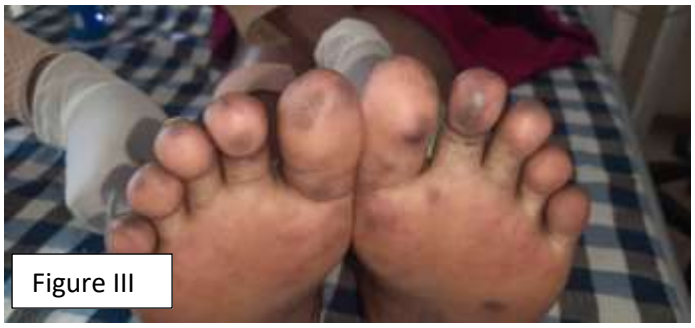

Figure III

**Discussion**

Vasculitis is an inflammation of the blood vessel wall which may have various clinical presentations depending on the size of the vessel involved (small, medium, and large) and site of the vessel (cutaneous or visceral). Prognosis may range from mild cases to severe and life threatening. SLE is a recognized cause of secondary vasculitis which may manifest in as many as 56% of SLE patients throughout their life. Cutaneous vasculitis is the most common form of SLE vasculitis (2, 3). Visceral involvement is identified as < 10% but can be life threatening and may need aggressive treatment.

There is a wide spectrum of presentation for cutaneous vasculitis ranging from lupus nonspecific lesions (purpura, petechiae, punctate vasculitic lesions, urticarial vasculitis, panniculitis, papulonodular lesions, or bullous lesions of the extremities, livedo reticularis, cutaneous infarction, erythematous plaques or macules, erythema with necrosis, panniculitis, splinter hemorrhages, and superficial ulcerations) to peripheral ischemia and gangrene (4). According to the literature patients with SLE vasculitis are mainly female and tend to be of young age. Our patient is a young female with first presentation as vasculitis. Visceral vasculitis in SLE usually occurs simultaneously with systemic flares usually in association with or following cutaneous vasculitis.

In two large cohort studies in patients with vasculitis and SLE, the most frequent type of vasculitis was leukocytoclastic vasculitis (60%), which was the type in our patient as well, followed by cryoglobulinemic vasculitis (25–30%) and urticarial vasculitis (7%) (4).

Antibodies against endothelial cells have been identified as a major factor in the pathogenesis of several connective tissue diseases, mainly vasculitidies. About 80% of SLE patients are positive for AECAs (anti endothelial cell antibodies). Drug induced and infection induced vasculitis are the other forms of SLE related vasculitis. Here, the vascular damage is caused via direct vessel wall involvement by the pathogens or via antigen induced autoimmune and inflammatory process. Our patient was previously healthy and was not on any long term or short term medications and her infection screening was negative.

Peripheral ischemia and gangrene has been described as a rare clinical entity in SLE. Prevalence of peripheral gangrene in SLE is 1.3 %( 5). Further, peripheral gangrene as first presentation of SLE is very rare and it is 0.2% in SLE patients and it is often found in the late stage of the disease (6). The underlying mechanism and pathophysiology of digital gangrene in SLE is thromboembolism, premature atherosclerosis, vasospasm, hypercoagulability and vasculitis (7).

According to the former literature, history of Raynaud's Phenomenon (RP) considered to be one of major predictive factors of peripheral gangrene in SLE which was not the case in our patient (7). RP can be seen in 50% of patients with SLE.

Secondary Anti Phospholipid Antibody Syndrome (APLS) is the other frequently described cause for the peripheral gangrene in SLE patients (7,8). Prevalence

of peripheral gangrene in APLS is 3.3 to 7.5%. APLS is characterized by positive antiphopspholipid antibodies (Anticardiolipin antibody, Anti-beta ii glycoprotein antibody and Lupus anticoagulant) which contribute to formation of thrombus. Clinical features of APLS include livedo reticularis, chronic ulcers, arterial or venous thrombosis, recurrent abortions and neurological symptoms. Although our patient had low positive Anticardiolipin antibodies, she didn't have any clinical features related to APLS. Further, antibody level should present in two or more occasions at least 12 weeks apart.

Accelerated atherosclerosis which usually occurs in long standing SLE is again a cause for peripheral ischemia and gangrene. Our patient had no history of dyslipidemia or any other risk factor for premature atherosclerosis. Her lipid profile and fasting blood sugar were within normal limits.

Other predictive factors include long standing history of SLE and elevated C-reactive proteins. In our patient, CRP was elevated but she had no previous history of SLE. A study by Liu et al in 2684 SLE patients, where 18 patients had digital gangrene and their average age at complication was 33.1±11.8 years and the average disease duration was 99.1±60.1 months. It was also observed that longer disease duration (≥4 years), RP and elevated CRP were highly prone for digital gangrene (9).

Underlying pathology of ischemia (vasculopathy, vasculitis, or vasospasm) will define management options such as vasodilator drugs which would be required in patients with RP. Early diagnosis and treatment including corticosteroid, immunosuppressive and lipid-lowering agents and anticoagulation are streamlined management strategies of digital gangrene in SLE. Corticosteroid reduces the risk of digital amputation.

According to the updated EULAR (European League Against Rheumatism) recommendations, lupus vasculitis treatment should be tailored to the severity of the disease and its associated symptoms. Mild-to-moderate manifestations are usually handled with oral corticosteroids and immunosuppressant such as methotrexate, azathioprine, and mycophenolate mofetil. A more aggressive approach with intravenous high-dose corticosteroids, cyclophosphamide, rituximab, intravenous immunoglobulin, and/or plasmapheresis is considered for the severe and life-threatening forms (4).

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# CHALLENGES IN MANAGING TYPE 1 DIABETES IN A TODDLER – A CASE REPORT

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## Abstract

Type 1 diabetes in toddlers and infants is a very challenging clinical entity for both patients and health care team. During this period of rapid physical and neurological development, they have distinct issues in diagnosis and management compared to older children. This case report describes a 2-year-old boy who was admitted with abdominal pain and increased frequency for 2 days. Diabetes ketoacidosis was confirmed by further evaluation. The acute clinical condition was resolved with appropriate fluid and insulin treatment. Following discharge from the hospital, his hyperglycaemia was very difficult to control. The challenges and struggles faced by the treating team and parents of a toddler with type 1 diabetes has been described here.

**Key words**: Type 1 diabetes, toddler, diabetes-ketoacidosis

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## Case History

A two-year boy was referred by a general physician to the paediatric unit with two days history of abdominal pain and increased urinary frequency. While investigating for possible urinary tract infection he was found to have glycosuria of 3+ which prompt the urgent referral. He was born at term and had normal developmental mile stones. His vaccination was up-to date. There was no other significant medical or family history. He was the only child and his parents were well educated.

His initial assessment revealed healthy looking child with mild dehydration and tachycardia. He had a height of 85cm and weight of 11kg ($25^{th}$ and $10^{th}$ centile in CHD). The boy was hyperglycaemic with plasma glucose of 370mg/dl and acidotic with PH of 7.23 and bicarbonate level of 3.8mmol/l. Urinalysis revealed glycosuria and ketonuria. Appropriate fluid resuscitation and insulin infusion resolved the diabetic ketoacidosis over 20 hours. His infection screening was negative. After 24 hours he was started with subcutaneous insulin therapy with soluble insulin 2-4 units thrice daily and isophane insulin 4 units at night which kept his pre-meal glucose value around 200mg/dl. Mother was educated on the diagnosis of T1DM and its chronic nature which requires lifelong commitment. Principles of diet and insulin therapy along with self-monitoring of glucose was discussed by the diabetes health care team. Later the child was discharged with premixed insulin analogs (30:70) 6 units mane and 4 units nocte with pen device. Follow-up visits were arranged as combine paediatric and endocrinology clinic.

On follow-up in 2 weeks mother had so many concerns to be discussed. She was quiet good with injections and self-monitoring. But glucose values are very much fluctuating and mostly high values above 250mg/dl. She was struggling on diet control and the child's preference over biscuits and sugary food. Understandably, she was very worried about the child's heath and possibility of T1DM in her future pregnancies. Her concerns were discussed and basics of management were reiterated. Follow-up visit with Nutritionist also arranged on the same day with a plan of arranging a review by paediatric endocrinology team at Lady Ridgeway hospital when current Covid situation has been settled.

**Discussion**

Among patients with Diabetes mellitus, 5 to 10% are having type 1 diabetes mellitus (T1DM). Worldwide, there are 600,900 children under 15 years of age are living with T1DM. In South East Asia the figures are around 184,000 [(1)]. Although children can develop T1DM at any age, a small minority of 2-4 % develop during infancy and toddlers' age [(2,3)]. Very young children pose significant challenge to the clinicians in diagnosing as well as managing this chronic illness. Majority may not present with the classic symptoms of diabetes, even if present could be missed. Polyuria and polydipsia may be considered normal by the parents when a child drinks plenty of fluids and urinate well. It is not un-common for children to present with respiratory or urinary tract infection or diaper candidal dermatitis. But compared to older children, toddlers are more prone to severe and rapid form of diabetes such as diabetes ketoacidosis, hyper-osmolality or even cerebral oedema. In addition, they have relatively low glycosylated haemoglobin and high titres of diabetes related autoantibodies at presentation indicating aggressive autoimmune insult [(3)]. This child was presented with diabetes keto-acidosis but luckily picked up early. Therefore, it is important to be vigilant and detect diabetes early which prevent the child form severe dehydration and shock.

Once the diagnosis has been made and the acute condition has been settled it is not easy to set-up an appropriate treatment regimen for these young children. When compared to older children, toddlers usually have high insulin requirement at initial period and short honeymoon period due to rapid pancreatic beta cell loss. Also they have extremely fluctuating glucose values. This could be due to number of factors which are unique to this age group [(4)]. As this is the crucial period of rapid growth spurt and neurological development, it is necessary to adjust the insulin and nutrition requirement frequently. Regular meal pattern is difficult among infants and toddlers; they often have erratic eating pattern or they take food every couple of hours. Similarly, food refusal also a major issue. They may not understand why they could not eat the similar foods that their friends or siblings eat. These young children quickly recognize parental stress on meal planning and use diabetes to bribe their favourite food. In the same way, physical activity which will improve glucose utilization, also much vary day to day for toddlers.

Furthermore, administering insulin to this young kids have number of practical issues. The difficulties of an active child to keep still for injections and glucose monitoring will increase parental anxiety. They are very sensitive to insulin and also have very unpredictable glucose values. The risk of hypoglycaemia is much severe and very challenging issue in the management. Very young children are

unable to communicate their symptoms of hypoglycaemia. Nocturnal hypoglycaemia is one of the worst problem due to prolonged overnight fasting. Effects of severe hypoglycaemia on the developing brain with regard to cognitive impairment is a significant issue in management.

Consideration of all the above factors are essential when managing an infant or toddler with diabetes. Generally, healthy food provided for normal infants would be acceptable if given regularly. Although input from an experienced dietician on infants and toddlers nutrition and diabetes care would clearly help parents on food choices and carbohydrate counting. Parents should be advised not to force feed or fight with the children on meal time; also not to drag the time for a long period on feeding. Generally, meals and snacks with complex carbohydrates should be encouraged and concentrated carbohydrates should be limited.

Toddlers are usually active, but their activities are variable. Physical activity must be encouraged as this plays an important role on their health in adolescents and adulthood. They need extra snack and may need glucose monitoring during additional activities.

Although children with T1DM are ideally treated with basal bolus insulin regimen via four or multiple injections, infants and toddlers with diabetes are usually given once or twice daily insulin injections. To avoid the risk of hypoglycaemia it would be better to start small dose of 0.25 – 0.5 units/kg body weight and slowly increasing by 1-2 units per day while monitoring the glucose values [(2)]. Long acting insulin analogs which are not licenced for very young kids could be considered due to their favourable pharmaco-dynamic profile. Their minimal peak action, slow and continuous systemic absorption and longer duration of action would be very convenient to use at-least during the initial period after diagnosis. Ultimately these children will be switched to multiple daily injections owing to their unpredictable glycaemic variability. More sensible approach would be administering rapid acting analogs after the meal to match the insulin dose to carbohydrate.

Self-monitoring of blood glucose using glucometer is a very useful tool in managing children with T1DM. Checking the glucose values before insulin injections will be useful in dose adjustments; 2 hour post prandial glucose and midnight values aid in predicting and preventing hypoglycaemia. Generally, blood glucose control in these young children involves less aggressive approach. Because of potential relationship between hypoglycaemia and later cognitive impairment the target HBA1c of less than 8% and less than 8.5% if excessive hypoglycaemia occurs is recommended [(4)]. Parents are usually advised to keep the pre-meal or pre-snack glucose value of 6 to 10 mmol/l. In addition parents should be educated on sick-day management since this age group is more vulnerable to get frequent respiratory and gastrointestinal infections.

The diagnosis of T1DM however change the daily activities of these young children including meals, play time, sleep, sibling and peer interaction and day care activities. This will substantially increase the psychological stress and alter the normal parenting experience. The daily responsibility of T1DM management and the worry of their child's health has potential interference with parental adjustment. This will reduce the quality of life of the children as well as the family members. Therefore, healthcare providers should not forget the parents' distress.

Discussing parents' concerns, coping skills and available support may help on their wellbeing. Specialised clinics meant for very young kids may allow parents to share their experiences and challenges which will improve their coping skills. Several benefits have been shown in behavioural interventions for parents and young children with T1DM [(5)].

**Conclusion**

Management of infants and toddlers with T1DM is distinct from older kids with diabetes. Biological, clinical and psychological issues related to this developmental period contributes so much struggle to the parents and healthcare providers in controlling diabetes while minimising family stress. Success of this difficult long journey depends on motivated team including family members, experienced diabetes health care team and supportive health system. In our set-up close cooperation between regional hospital where the affected child lives and teaching hospital where specialised diabetes care team is available should be encouraged.

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Conflict of interest: None

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# NON-TRAUMATIC DEPRESSED SKULL FRACTURE WITH EPIDURAL HAEMORRHAGE IN A NEONATE

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## Abstract

This case study describes the finding of a depressed skull fracture causing epidural haemorrhage in a neonate who was delivered without instrumentation and with no history of trauma. Instrumental deliveries typically cause skull fractures in newborns. Spontaneous skull fractures are rare but can happen due to trauma even during vaginal delivery. (1) Epidural hematoma (EDH) is rare and always post-traumatic in origin. (2) The imaging of choice in diagnosing acute EDH is Computed Tomography (CT), though initial surveillance can be done using an Ultra Sound Scan (USS). (3) The overall prognosis for a child born with a skull fracture is good. (4)

**Key Words:** Neonate, Epidural haemorrhage, Skull fracture

## Introduction

Instrumental deliveries usually cause skull fractures in newborns. We report a case of a spontaneously delivered neonate who presented with convulsions on day one with a depressed skull fracture associated with EDH. Computer Tomography (CT) was the imaging of choice in diagnosing acute Epidural Haemorrhage (EDH). (3)

## Case History

A baby girl was born at POA 37+4 with a birth weight of 3 Kg and delivered vaginally without using instruments. At birth, she had a cephalhaematoma at the posterior aspect. The baby developed a convulsion at 15 hours. An urgent Ultra Sound Scan brain was performed to exclude intracranial pathology.

USS brain demonstrated an extra-axial hypoechoic region in the left parietal region, indicating extra-axial haemorrhage suggesting acute subdural haemorrhage (SDH) (Image-01). Colour Doppler parameters revealed reduced flow in the superior sagittal sinus and normal intracranial arteries. CT venogram was performed to exclude venous sinus thrombosis and better characterize the hemorrhage. It showed a left parietal extra-axial biconvex hyperdense area measuring 6.5mm in thickness associated with subgaleal haematoma with a depressed skull fracture (Image -02). The venogram was unremarkable. As there was no clear history of trauma to exclude bone fragility disorder, she underwent a full body X-ray with no evidence of fractures elsewhere.

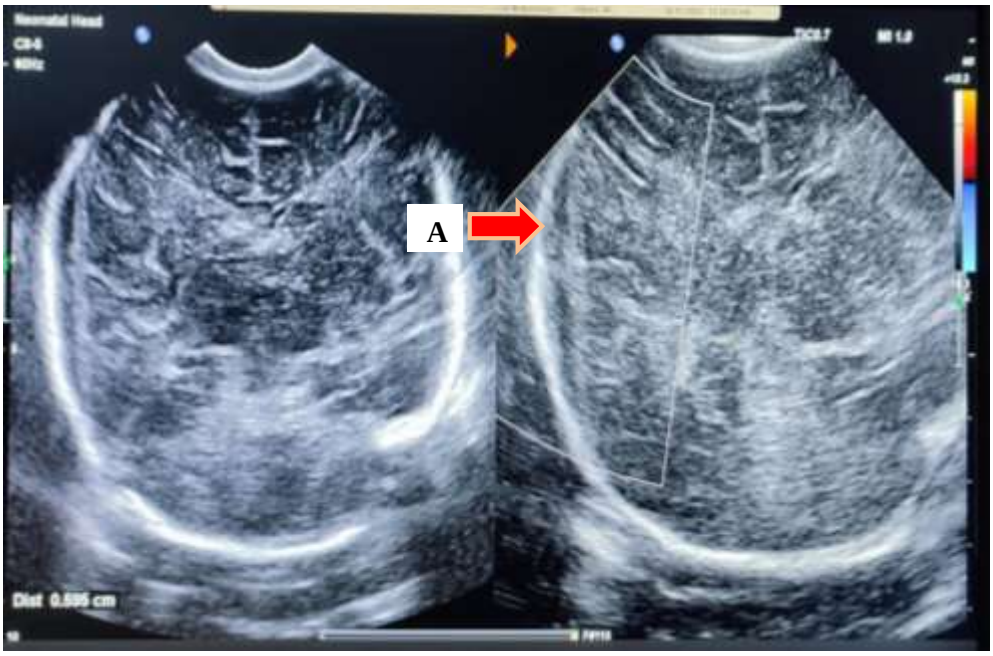


Image 1- Axial USS image of the brain with left parietal extra-axial hemorrhage Arrow A

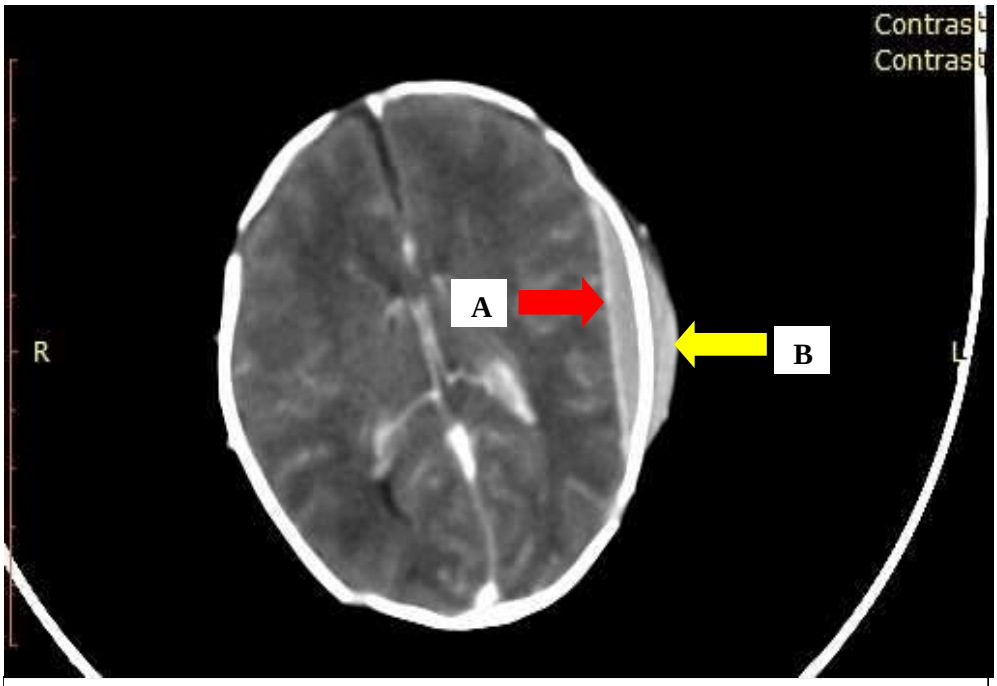


Image 2- Axial contrast CT brain image showing left Parietal extra-axial and subgaleal hematoma Arrow A Arrow B

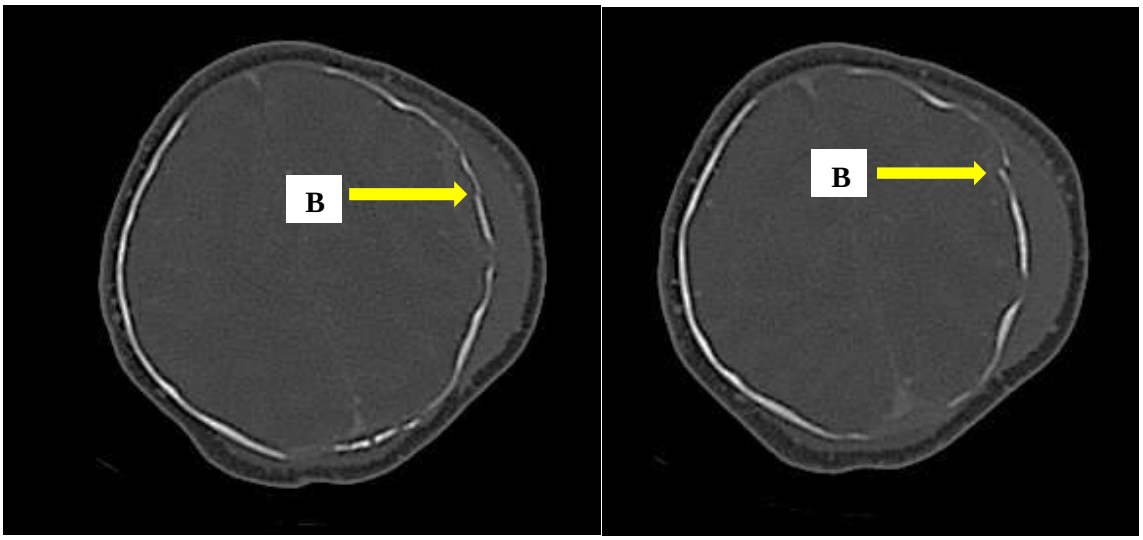


Figure 3- Axial Contrast CT bone window showing the depressed skull fracture Arrow B

Left parietal EDH evacuation was performed via burr hole under general anesthesia. The follow-up CT scan shows pneumocephalus at the surgical site of EDH evacuation. The baby was discharged four days after evacuation with anti-epileptic drugs, and planned to tail off drugs and continue to follow up.

**Discussion**

Skull fractures in newborns are usually caused by instrumental deliveries using forceps, vacuum devices or due to mechanical forces applied assisting delivery in nulliparous women with a difficult delivery. [(1) (5)] Spontaneous skull fractures are rare but still possible, which is the cause in our case. [(5) (6)] During vaginal delivery, the fetal head gets compressed by the mother's pelvic bony structures, which might lead to skull fractures and brain injuries in newborns. [(1) (2) (5)] A depressed skull fracture is one such fracture that causes part of the skull bone to sink inwards and thus has the potential to be more severe since it is more likely to cause bleeding. [(4)] Acute epidural hematoma (EDH) is a type of intracranial bleed that can associate with skull fractures. EDH also could be associated with instrument delivery, coagulopathy, or fragile bone disease in a neonate. [(7) (4)]

An ultrasound scan of the brain is indicated for all neonates with risk or suspicion of brain pathology, as in our case, who presented with convulsions. [(8)] In our patient, differentiation between EDH and SDH was difficult due to the fracture's non-visualization and the lesion's crescentic shape. EDH in the USS brain shows well defined lentiform /biconvex shape hypoechoic region just beneath the skull, and an extra-axial crescentic hypoechoic area can be seen in SDH. [(9)] 11% of EDH in children cross sutures due to skull fracture across the sutures, favouring SDH. Suture diastasis and vertex extra dural haematoma can also favour SDH. [(3) (10)(11)]

USS brain can be used as a screening investigation to detect skull fracture but has a chance of false negative results, similar to our case in which skull fracture was revealed neither in the USS brain nor clinically (due to overlapping cephalhaematoma). [(12)] Dural venous sinus thrombosis is a known complication of skull

fracture. [13] As in our case, CT brain is critical to confirm and rule out the diagnosis of intracranial haemorrhage and follow up after surgical evacuation.

Most of the time, these babies do not experience severe complications, and the overall prognosis for a child born with a skull fracture is good. [1] [2]

## Conclusion

Depressed skull fracture associated with EDH is a rare occurrence in spontaneous normal vaginal delivery without using instruments. However, it is still possible and vigilant attention to these babies helps in early diagnosis and helps timely intervention. Cranial USS is the first line imaging modality in neonates to identify cerebral pathology due to its availability and safety. CT is the gold standard image modality in traumatic intracranial hemorrhage.

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# STORY BEHIND A GRANULOMATOUS FACIAL LESION - ANGIOSARCOMA THE HIDDEN MONSTER

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## Abstract

Cutaneous angiosarcoma is rare and highly aggressive tumour of skin and soft tissue with the predilection to white skinned elderly. Given its variable clinical presentation it can mimic several clinical entities making the diagnosis difficult. Early diagnosis is important for its prognosis and will confirmed by histopathology and immunohistochemistry upon clinical suspicion. Herein, we report a case of cutaneous angiosarcoma presenting as a facial granulomatous plaque.

**Key Words** Cutaneous angiosarcoma, Facial, Granuloma

## Introduction

Infiltrative (Granulomatous) facial lesions are a well-known entity of presentation in a dermatology clinic. Underlying aetiology needs to be rule out using appropriate investigations excluding the clinical differential diagnoses. Common aetiologies of infiltrative (granulomatous) facial lesion are infectious diseases including leprosy, deep fungal infections, leishmaniasis and inflammatory conditions i.e. Wagener's granulomatosis and sarcoidosis. Angiosarcoma a highly aggressive tumour and presenting as a perinasal granulomatous plaque is not common. Thus we report a case of angiosarcoma presenting as a perinasal granulomatous plaque.

## Case History

A 75-year-old male with a background history of poorly controlled type 2 diabetes presented to dermatology department with asymptomatic perinasal skin lesion for two months duration. Other than the disfigurement he did not reveal any sinister symptom on systemic inquiry.

On examination, there was a poorly defined, erythematous, indurated, firm to hard plaque with irregular borders and irregular surface without any epidermal changes (Fig: 1) and the lesion was negative for apple-jelly colur on diascopic examination. There were no similar lesions elsewhere in the body and systemic examination was normal including regional lymph node examination.

His investigations revealed normal haematological parameters including normal ESR, normal renal and liver function. His chest X ray and Mantoux (5 mm) were normal, slit skin smears for AFB and Geimsa were negative, TB PCR and leishmanial PCR also were negative.

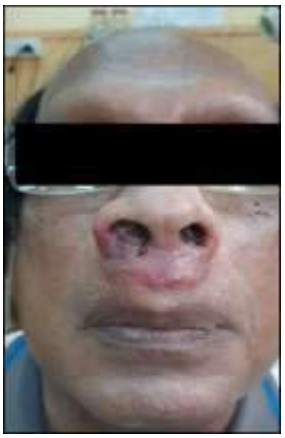
The histopathology revealed only a chronic inflammatory cell infiltrate with possible early granuloma formation. The special stains which were negative include grocott and fite stain. Tissue cultures for tuberculosis/leishmania/fungi were negative.

Since the patient was getting symptomatic with local obstructive features in the half way through investigations (Figure-02) and the first biopsy didn't reveal any diagnostic

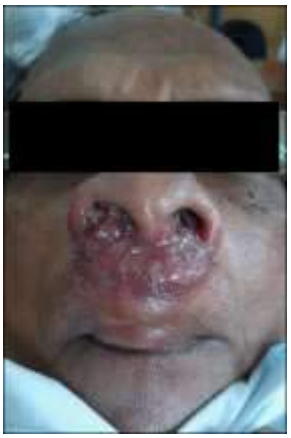
clue a second, deep, incisional biopsy was performed.

The histology of it showed skin with a vascular lesion with complex anastomosing vascular channels lined by atypical endothelial cells (Figure -04) suggestive of an angiosarcoma.

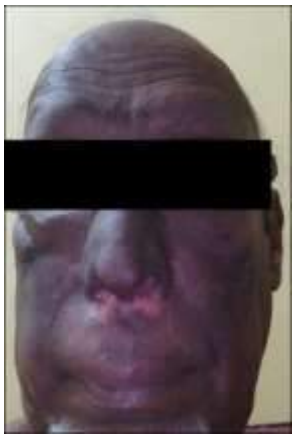
Then, it was confirmed with immunohistochemical studies with strongly positive for CD 31 (Figure-05) and vimentin (Figure-06),SMA and negative for CD 45, desmin,Myo D 1,Melan A and HMB 45 which confirmed the diagnosis of high grade angiosarcoma. The patient was referred to Oncology unit. Currently the patient is on chemotherapy and radiotherapy to which the tumour has responded (Figure -03).



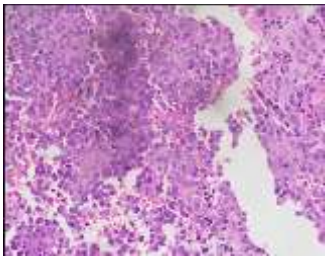
(Fig: 1) Initial presentation



(Fig: 2) after one month from Initial presentation



(Fig: 3) after treatment



(Fig: 4) H&E section

(Fig: 5) CD 31 positive stain

(Fig: 6) Vimentin positive stain

## Discussion

Facial infiltrated or granulomatous lesions are not very rare in a dermatology clinic. Clinical differential diagnoses are vital to plan out subsequent investigations. Histopathological and microbiological studies are pivotal in the process of making a complete diagnosis. Perinasal granulomatous lesion has its own set of differential diagnoses that a dermatologist tends to consider at a glance including sarcoidosis, leprosy, cutaneous tuberculosis and mucocutaneous leishmaniasis (1). Being a tropical country, the above differentials should be the first priority however some rare entities such as angiosarcomas and other malignant neoplasms should be considered in the differential diagnoses as

they may mimic granulomatous facial lesions as in our case and also the prognosis depends on the early diagnosis and early treatment.

Angiosarcomas are highly aggressive tumour of skin and soft tissue with five-year survival of less than 35% (1,2). Clinical presentation of cutaneous angiosarcoma of head and neck of the elderly include bruise-like lesion, violaceous nodules and plaques, and flat infiltrating haemorrhagic areas. Often these presentations can mimic several other conditions such as vascular malformations, nodular melanoma, lymphoma, sarcoidosis, or facial granuloma (2).

Clinical classification of Angio-Sarcoma (AS) includes;

1. AS on the face and scalp of elderly population – most common type,
2. AS either in association with chronic lymphedema or secondary to a prior surgery (Stewart-Treves syndrome),
3. AS following chronic radio dermatitis or skin trauma and ulceration(3).

Fast spreading nature with the highest rate of lymph node metastasis among all soft tissue sarcomas of the head and neck will account for the poor prognosis.

Histopathological features of angiosarcoma also variable as clinical features. The well-differentiated lesions may mimic hemangioma or lymphangioma, whereas the poorly differentiated lesions mimic melanoma(4). In such situations, immunostaining for markers will help in the diagnosis without any delays.

Management options include surgical resection, radiotherapy and chemotherapy which has to be decided on the grounds of tumour bulk, lymph node involvement and patient co-morbidities.

In the context of varying clinical presentations of angiosarcoma it should also include in the clinical differential diagnoses of infiltrative skin lesions particularly head and neck area in elderly patients.

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# SIBLINGS DIAGNOSED WITH PAPILLON-LAFEVRE SYNDROME; ONE PRESENTED WITH RECURRENT LIVER ABSCESS; THE OTHER PRESENTED WITH HYPERKERATOSIS AND SKIN INFECTION

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## Abstract

The Papillon-Lafevre syndrome is an extremely rare autosomal recessive disorder has affected about 1-4 per million people globally. The cathepsin C enzyme activity is deficient due to cathepsin C gene mutation in these patients and characteristic features are palmoplantar hyperkeratosis, loss of deciduous and permanent teeth and increased susceptibility to infections. The palmoplantar hyperkeratosis and periodontopathy usually start before age of 4 years. Here we present two cases of Papillon-Lafevre syndrome of two siblings, brother and a sister. A 6 year old boy with a history of recurrent liver abscesses presented with liver abscess again and with palmoplantar hyperkeratosis and already teeth loss. A 12 year old girl who presented with skin infection was also found to have hyperkeratosis and lost her teeth as well. Both were clinically and genetically confirmed to have Papillon-Lafevre syndrome. Early identification and early treatment may improve the outcome of the management of patients with Papillon-Lafevre syndrome.

**Keywords:** Papillon-Lafevre syndrome, Cathepsin C gene, Palmoplantar hyperkeratosis, Periodontopathy

## Case History

A 6 year old boy presented to pediatric ward at DGH-Nuwara Eliya with the history of fever for 7 days duration with a background of recurrent abscess formation including right lobe of liver in 10/2020, right groin in 05/2021, another liver abscess in 08/2021, and an abdominal wall abscess in 08/2021 which were initially managed by the surgical team. The child was born at term via uncomplicated vaginal delivery with birth weight of 3.65kg. Postnatal period was uncomplicated and vaccination was up to date. His growth was following the centiles and development was normal. He has two siblings, middle one was healthy and elder one also diagnosed as Papillon-Lefevre syndrome later. He has no family history of immunodeficiency.

On examination, child was active & alert, cachectic, not pale, anicteric, hyperkeratosis over the palms and soles was noted he did not have clubbing, flapping tremors or spider naïve. He had lost his teeth already. He had a heart rate of 120 beats per minute with blood pressure of 90/60 mmHg,

abdomen soft and non-tender with no palpable organomegaly, bilateral lungs had equal air entry. Child was directed to regular pediatric clinic follow-up but again child appeared with complaining of poorly resolving fever with simple antipyretics for 7 days duration with loss of appetite. The fever associated with chills and rigors but there were no other clinical symptoms and signs. His full blood count showed neutrophil leukocytosis with high platelets and hemoglobin was 7.8 g/dL. His CRP was high. Other blood investigations were normal. Ultrasound scan (USS) of the abdomen revealed 6.5×4.7×8.5 cm size liver abscess of segment vii breaching the capsule and continuing into right sub phrenic area forming right sub diaphragmatic collection. Abscess contained organized material and no drainable liquid material seen. Right diaphragm appears intact however reactive right basal effusion with consolidation seen in right lung base. His Immunoglobulin levels are IgG >3000(559-1492), IgA - 346(54-221), IgM -234(27-118), IgE > 2000(1-411).

Child was started on IV Cefotaxime 750mg 8 hourly, IV Vancomycin 225mg 8 hourly, IV Metronidazole 120mg 8 hourly and oral Fluconazole 100mg daily. After 10 days of IV antibiotics abscess was drained under USS guidance. A course of 28 days of IV Cefotaxime, 24 days of IV Vancomycine and 12 days of metronidazole was given. The cytology of liver abscess showed necrotic tissues only and GENE X- pert was negative for tuberculosis. Child was discharged from the ward with a plan to readmit to pediatric ward after one week to reassess the abscess and to arrange genetic assays. Genetic assay revealed two pathogenic variant in CTSC. CTSC is associated with autosomal recessive Papillon-Lefevre syndrome (PLS) and related disorders.

A 12 year old girl, the elder sister of the above child who already has had hyperkeratosis has presented to the pediatric ward at DGH- Nuwara Eliya due to worsening of rash. She has lost her teeth but she had no history of liver abscess formation prior or other significant past medical history other than the rash. Her birth, growth and development history were unremarkable. She was diagnosed as pyogenic skin infection and managed with IV antibiotics for 7 days duration. Later she was genetically confirmed of Papillon-Lafevre syndrome. Dermatological and orthodontist follow up were arranged and 10 months later she developed liver abscess which was initially managed with IV antibiotics for about 7 days and then aspiration done bwith USS-guidance. Due to poor response to IV antibiotics the USS-guided drain was inserted to the abscess and removed after 6 days. The abscess aspirate culture was positive for Methicillin Resistant Staphylococcus Aureus (MRSA). She was managed with IV antibiotics for one month duration and discharged with follow up arrangement.

**Discussion**

The Papillon-Lafevre syndrome (PL) is an autosomal recessive disease and is extremely rare.[1] The prevalence of PL is about 1-4 case per million globally.[1] The mutation of the gene lies on chromosome (11q14-q21) leads to total absence of

cathepsin C (CTCS) activity , a lysosomal protease.[2] This gene is mainly expressed in epithelial regions like palms, soles, gingiva and immune cells and their precursors. [2]

The clinical features of PL syndrome include palmoplantar keratoderma which usually starts between 1-4 years and extends trans gradient pattern, periodontitis which is usually start at 3-4 years of age and transverse grooving and ridging of nails. In our cases, both had palmoplantar keratoderma and periodontitis but none of them had nail changes. [3] The susceptibility of infection due to immune system dysfunction is about 20% among PL patients.[4] In our cases the boy had recurrent liver abscess and abdominal wall abscess including the groin while the girl had skin infection and later admitted with liver abscess. Intracranial calcification of choroid plexus and tentorium as well as palmoplantar hyperhidrosis are the other features of PL.[4]

There are two main differential diagnoses(DD) for PL including Haim-Munk Syndrome which is an allelic variant of PL and in those patients other than the palmoplantar hyperkeratosis and loss of dentition, arachnodactyly and acroosteolysis can be seen.[5] Other DD is Prepubertal Periodontitis where palmoplantar hyperkeratosis is absent.[6]

The main treatment modality in PL is the oral retinoid like acitretin, etretinate and isotretinion with the combination of antimicrobial treatment.[7] The periodontitis improves with the oral hygienic measures and extraction of primary teeth with combination of oral antibiotics. [8]

## Conclusion

The PL Syndrome is extremely rare in the world and further rarer in Sri Lanka as well. The early identification might protect them from losing permanent teeth and can take early precautions to prevent infections. These children need proper follow-up and sometimes need antimicrobial prophylaxis for some duration after having infection.

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# INCIDENTALLY FOUND MESENTERIC PSEUDOCYST OF THE ILEUM

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## Abstract

Mesenteric cyst, a relatively rare tumour originating from the mesentery or the surrounding area is located in the mesentery of the small intestine and mesocolon in most cases. They are frequently discovered incidentally with Ultra Sound Scan (USS) or computed tomography (CT) as they are asymptomatic. They are either simple or multiple and either unilocular or multilocular and they may contain hemorrhagic, serous, chylous or infected fluid. Various complications have been associated with mesenteric cysts including intestinal obstruction, volvulous, haemorrhage into the cyst, infection, rupture, cystic torsion, obstruction of the urinary and biliary tract. The most common indication for surgical removal is the presence of an abdominal mass. The goal of surgical therapy is complete excision. We present a case of 3 year old boy with abdominal pain who found to have mesenteric cyst intra-operatively and excised completely.

**Key Words**: Mesenteric cysts, Intestinal obstruction, Loculation.

## Introduction

Mesenteric cyst is a relatively rare tumour arising within the abdominal cavity that originate from the mesentery or the surrounding area and has no linkage with retroperitoneal organs. Although it may occur in any part of the mesentery it is located in the mesentery of the small intestine and mesocolon in most cases.(1)

Mesenteric cysts are frequently discovered incidentally with Ultra Sound Scan or computed tomography as they are asymptomatic. Some patients report symptoms associated with compression or complications of the cysts. Although USS and CT are the best modalities for accurate diagnosis prior to surgery, because of the origin of the lesion or the characteristics of the internal aspect of the cysts, they cannot be easily determined by radiological examination alone. (2)

We herein present a case of incidental finding of a mesenteric pseudocyst of the ileum intraoperatively in a 3-year-old boy who was suspected to be having acute appendicitis.

The patient underwent ileal resection and end to end ileal anastomosis.

## Case History

A 3-year-old boy who had been healthy previously, was admitted to the pediatric ward with a history of fever, abdominal pain, dysuria, reduced activity, and poor feeding for 3 days duration. No history of similar episodes of symptoms in the past. The child had a few episodes of loose stools for few days prior to admiossion. His past medical and past surgical history is unremarkable, and his growth and development is age appropriate.

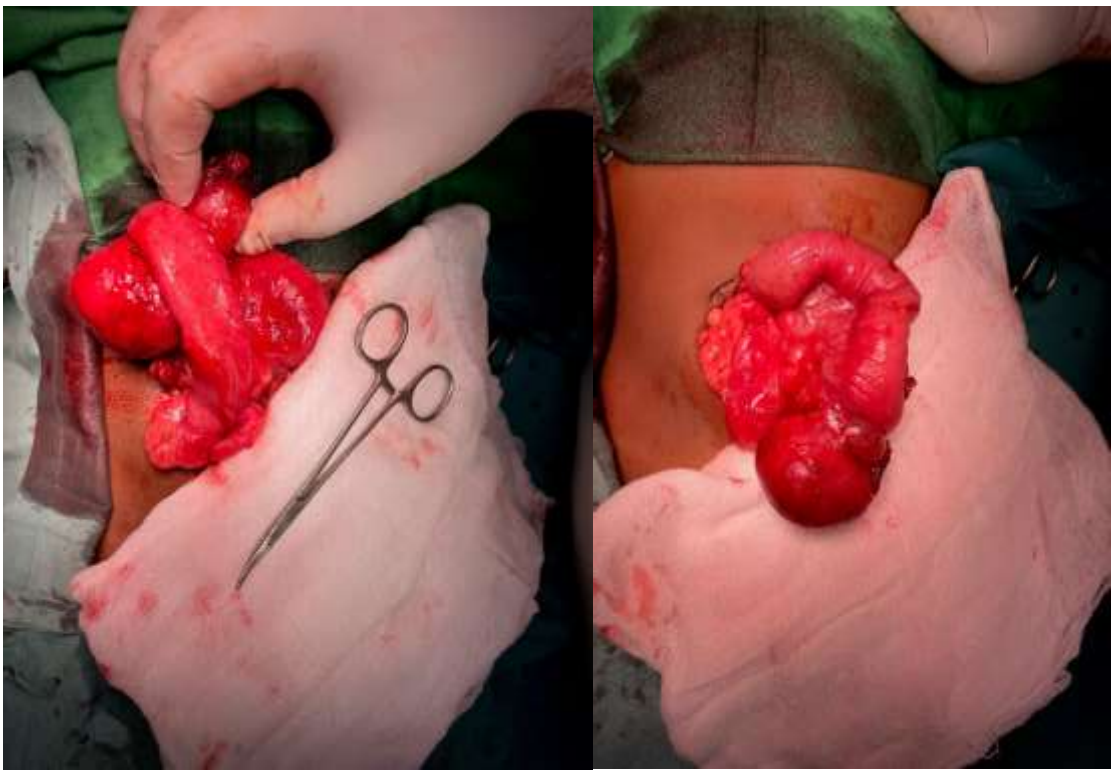
On examination child was ill-looking and febrile. On abdominal examination, there was significant right iliac fossa tenderness with

rebound tenderness as well. Examination of the systems was found to be unremarkable and the child was hemodynamically stable.
His inflammatory markers were found to be significantly elevated with marked neutrophil leukocytosis (WBC 20000, NEUT 11000) and a CRP was 114mg /dl. USS showed spherical-shaped septated collection in the right iliac fossa region extending towards the liver. The wall of the collection showed hyperemia with surrounding inflammatory changes indicating acute inflammation which arose the suspicion of a ruptured appendix forming an appendicular abscess.

An exploratory laparotomy had been performed and found to have a bifurcating cystic lesion of about 4*4cm in size on the enteric side of the ileum attached to the wall. There were multiple enlarged lymph nodes in the mesentery (Figure-1). The appendix and caecum appeared normal. Liver and rest of the bowel appeared normal. There was no peritoneal collection or abscess formation.
Small bowel segment of 10cm with the cyst had been resected and sent for histology. An end-to-end small bowel anastomosis was done.

His postoperative recovery was uneventful and the child was discharged on the fifth postoperative day.
Histology revealed mesenteric pseudocyst of the ileum. Macroscopically the cystic lesion is attached to the ileal wall measuring 60*40*30mm.It was filled with mucinous material. Microscopy showed ileal tissue with a cyst in the sub-serosa.

*Figure 1- Intraoperative finding of mesenteric pseudocyst of the ileum*



## Discussion

Mesenteric cyst is a rare condition with the incidence of about 1 per 140,000 general hospital admissions and about 1 per 20,000 pediatric hospital admissions. Approximately 1/3 of cases occur in children younger than 15 years. [(2)]

Mesenteric cysts most commonly occur in the small bowel mesentery on the mesenteric side of the bowel. They are either simple or multiple and either unilocular or multilocular and they may contain hemorrhagic, serous, chylous or infected fluid. The fluid is serous in ileal and colonic cysts and is chylous in jejunal cysts. [(3)]

Mesenteric cysts are thought to represent the benign proliferations of ectopic lymphatics that lack communication with the normal lymphatic system. Cysts are thought to arise from lymphatic spaces associated with the embryonic retroperitoneal lymph sac.

Various complications have been associated with mesenteric cysts including intestinal obstruction, volvulous, haemorrhage into the cyst, infection, rupture, cystic torsion, obstruction of the urinary and biliary tract. Malignant transformation of mesenteric cysts has been reported in adults, but not in children. [(4)]

No medical treatment is available. The most common indication for surgical intervention is the presence of an abdominal mass with or without signs of intestinal obstruction.[(5)] The goal of surgical therapy is complete excision.

## Conclusion

Though the entity of the mesenteric cyst is very rare, early suspicion and diagnosis with complete removal surgically will avoid the subsequent complications and reduce the morbidity and mortality.

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# SUCCESSFUL DRAINAGE OF RETROPHARYNGEAL AND PARAPHARYNGEAL ABSCESS IN A CHILD WITH USS GUIDANCE- A CASE REPORT

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## Abstract

Deep neck abscesses in children are not so common and they can present with minimal symptoms especially in younger children. Thus need to be evaluated carefully for the diagnosis. We share a case of a 2 months old otherwise healthy child present with non-specific symptoms for four days duration. Ultrasound scan (USS) of the neck revealed a large retro-pharyngeal and para-pharyngeal abscess measuring 3.5× 2 ×4 cm which was successfully drained with USS guidance. *Staphylococcus aureus* was isolated and treated with targeted intravenous antibiotics for 7 days. Child was fully recovered and kept under surveillance for 2 months and discharged from follow up.

**Key words**: Pharyngeal abscess, Guided drainage, *Staphylococcus aureus*

## Case History

We present a case of retro-pharyngeal and para-pharyngeal abscess in a 2 month old baby boy who presented to pediatric ward at DGH Nuwara Eliya with nasal congestion and yellowish sputum production with difficulty in breathing for four days duration. He had no cough or fever but had a history of associated snoring. His birth and post natal histories were insignificant. He was growing along the centile and development was age appropriated. Immunization was up to date.

On examination, child was active, alert and afebrile. A mild stridor was noted but was not distressed. Respiratory rate was 36/minute and bilateral lungs were clear with equal air entry and haemodynamically stable. His full blood count showed neutrophil leukocytosis with high platelet count and hemoglobin was normal. Lateral neck x-ray revealed widening of the cervical prevertebral space in the neck suggestive of retropharyngeal abscess (Image-1), warranting an USS. USS demonstrated a large collection in retro-pharyngeal and para-pharyngeal spaces measuring 3.5× 2 ×4 cm (Image-2). Thyroid gland and vascular bundle appeared normal and there was no cervical lymphadenopathy. Computed tomography (CT) was not attempted due to Covid positive status and adequate visualization in the USS.

Pediatric ENT surgical opinion was taken and suggested an USS guided drainage of the abscess. Prior to guided drainage child was tested for Covid-19 with rapid antigen test it revealed a positive result. Ultrasound guided aspiration done under aseptic condition and strict covid-19 precautions by the pediatrician and the radiologist. Under USS guidance, 10 ml of pus drained out with 18 G needle without any post procedure complications. Second USS guided aspiration done and 5 ml of pus was drained on the following day. Culture of the aspirate isolated *staphylococcus aureus* after 24 hours. He was treated with intravenous Cefotaxime 250 mg 6 hourly, intravenous C.Penicillin 250mg 6 hourly, intravenous Teicoplanin 50 mg for 7 days according tosenitivity pattern and nebulization with budesonide and symptomatic treatment. Child was followed up for 2 months and discharged from follow up.

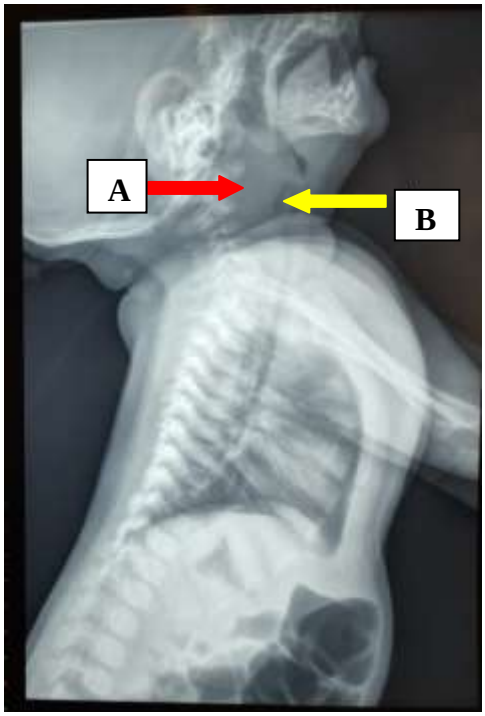


*Figure 1 - Lateral neck x-ray demonstrating widening of prevertebral space (Arrow A). The airway is indicated by the Arrow B.*

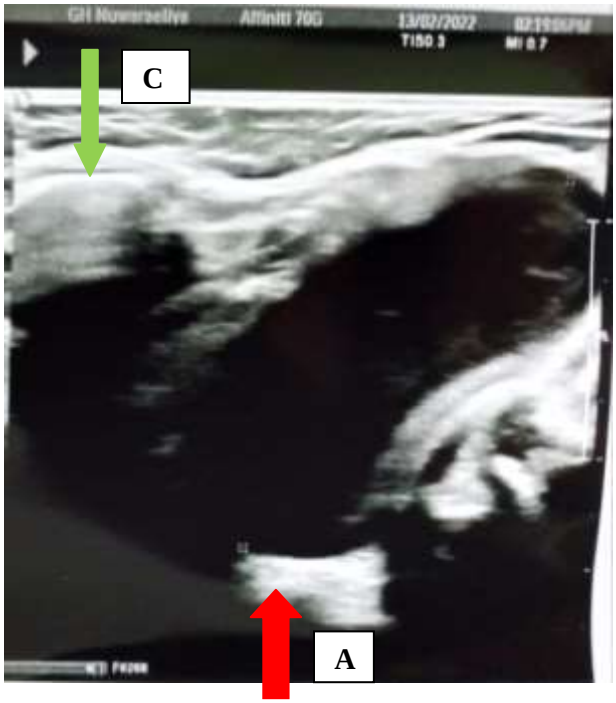


*Figure 2 - USS image of the neck demonstrating the Para pharyngeal abscess (Arrow A). The airway is indicated by the Arrow C.*

**Discussion**

The clinical presentation of retro-pharyngeal abscess in children is subtle and is uncommon when compared to adults. Children are unable to verbalize their symptoms and poor co-operation with examination make the diagnosis difficult.[1 2] It is more common among children under 6 years.[3] Common causes of retro-pharyngeal abscess in children are upper respiratory tract infection (67%), odontogenic infection (25.3%) and trauma and foreign body ingestion are less common with compared to adults.[3]

The space between the bucco-pharyngeal fascia which covers the posterior pharynx and the esophagus and the alar fascia is the retropharyngeal space. The inferior part of this space fused in the midline and contains two chains of lymph nodes. The extension of the space is from base of the skull to mediastinum.[3] The inverted pyramid shape space extending from the base of the skull to hyoid bone is the para-pharyngeal space.[3] Retro-pharyngeal abscesses are primarily seen early in childhood because the lymph nodes in the space tend to regress with age.[3] Retro-pharyngeal abscess mainly presents with neck pain and swelling, fever, irritability, dysphagia, dyspnea and drooling of saliva whereas para-pharyngeal abscesses present with fewer symptoms and therefore more difficult to diagnose.[3]

The common organisms causing deep neck abscesses are aerobic organisms like *staphylococcus aureus*, *staphylococcus viridans*, beta hemolytic streptococcus, gram negative organisms and anaerobic organisms.[2,3] The community acquired methicillin resistant S. aureus have been reported as high as 61% as a rising infection isolated in children with deep neck abscesses.[5] The main stay of empirical antibiotics are the Penicillin with combination of beta-lactamase inhibitor or beta-lactamase resistant antibiotic (Cefuroxime , Imipenum) together with anaerobic cover (metronidazole, clindamycin).[3] However culture directed therapy with clinical response would be helpful in choosing appropriate antibiotic.[1, 3]

CT may not be specific in diagnosis of neck abscess but has a sensitivity of 92%.[1, 3] In most cases CT is needed to visualize the extension. The lateral neck x- ray has a sensitivity of 83% and with the combination of positive neck x- ray and the clinical findings are more in favor of the diagnosis.[3] The ultrasound scan of neck has a more accurate role in differentiating a drainable abscess from cellulitis with compared to CT.[3] In our case USS played a major role and CT was not performed. Though the Magnetic Resonance Imaging (MRI) is the best to view the soft tissues considering the time taken for the procedure it is not usually performed.[3]Furthermore, MRI is not readily available, and may need general anaesthesia in infants, restricting its use in acute setting.

The smaller abscess (<2.2cm) in older children (>4 years) can be managed conservatively with antibiotics but close observation is essential and threshold to operate should be low because of the risk of mediastinitis without much clinical findings.[1,3] The large abscess should be managed with surgically.[1,3] An external approach is commonly used to drain uncomplicated abscesses or when there is medial displacement of adjacent structures. An intra-oral approach is more safer to avoid neuro-vascular injury.[3] If the air way is not compromised minimally invasive techniques like image guided needle aspiration can be used.[3,4] In the index child we used USS- guided drainage with success.

Complications of deep neck abscess includes mediastinitis, internal jugular vein thrombo-phlebitis, carotid artery aneurysm, necrotizing fasciitis but all can be prevented

by early identification and appropriate treatment.[3]

## Conclusion

Evaluation should be more careful in children to diagnose retropharyngeal and parapharyngeal abscesses as they are uncommon and the presentation is subtle. The gram positives are the most common organisms while the gram negative and anaerobic is unable to be removed from the picture. The MRSA cases are increasing in number as well.

Though the MRI is best to see the soft tissues, considering time and availability it is out of passion in diagnosis of neck abscesses while ultrasound scan plays a good role in diagnosis as well as in management. CT scan might need specially if planning for surgery.

Thorough clinical evaluation, early diagnosis and early, targeted management will prevent serious complications of these abscesses.

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# CORONARY ARTERY FISTULAE: CARDIAC COMPUTED TOMOGRAPHY (CT) AS AN EXCELLENT IMAGING MODALITY; A REVIEW OF 2 CASES

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## Abstract

Coronary Artery Fistulae (CAFs) are abnormal connections of the coronary arteries that bypass the myocardial capillary bed and terminate into chambers of the heart or major blood vessels [1]. CAFs are rare, and most of them are congenital in origin. As CAFs can be asymptomatic and detected incidentally, the true incidence is difficult to evaluate [2].

CAFs usually have various and complicated anatomical features, and the clinical symptoms mainly depend on the size, origin and drainage site of the fistulae. Thus, accurate imaging assessment of these characteristics is crucial for the diagnosis and plan out the management [2].

Due to the high temporal and spatial resolution, coronary CT angiography has recently become more widely used in cardiovascular disease diagnosis, and many asymptomatic CAFs are accidentally found. Here, we reviewed the imaging features of CAFs on coronary CT angiography, mainly focusing on the evaluation of anatomical features which enable in subsequent management [2].

## Introduction

An abnormal connection between an epicardial coronary artery into a cardiac chamber, great vessel, or other vascular structure (mediastinal vessels, coronary sinus) is known as a coronary artery fistula [1]. These anomalies can be congenital or acquired in origin; they can be presented at birth or be observed later in life. Congenital fistulae results from failure of fetal structures such as splanchnic veins or intramyocardial trabecular sinusoids to regress. Fistulae may be associated with other congenital abnormalities, including atrial or ventricular septal defects, patent ductus arteriosus, pulmonary atresia and Tetralogy of Fallot [1]. Acquired fistulae can result from intracardiac operations, trauma, and transcatheter procedures as a result of abnormal healing between the damaged circulatory chambers and vessels.

Fistulae that connect the left circulation to left-sided chambers may not result in clinical disease and be incidentally found on angiography. However, some will raise diastolic pressure in the affected chamber causing dilatation or hypertrophy. Fistulae between the left and right circulation result in shunt of blood flow, which may be clinically relevant, depending on the volume of flow [1].

**Case 1:**

A 48-year-old female presented with exertional dyspnea for 6 months duration. Her physical examination was unremarkable. Her Electrocardiogram( ECG) and chest X-ray were normal. A Transthoracic echocardiogram showed no significant abnormalities. Her CT Coronary Calcium Score was 126 AU. CT Coronary Angiogram (CA) revealed a coronary artery fistulous tract which is originating from the Left Coronary Sinus.

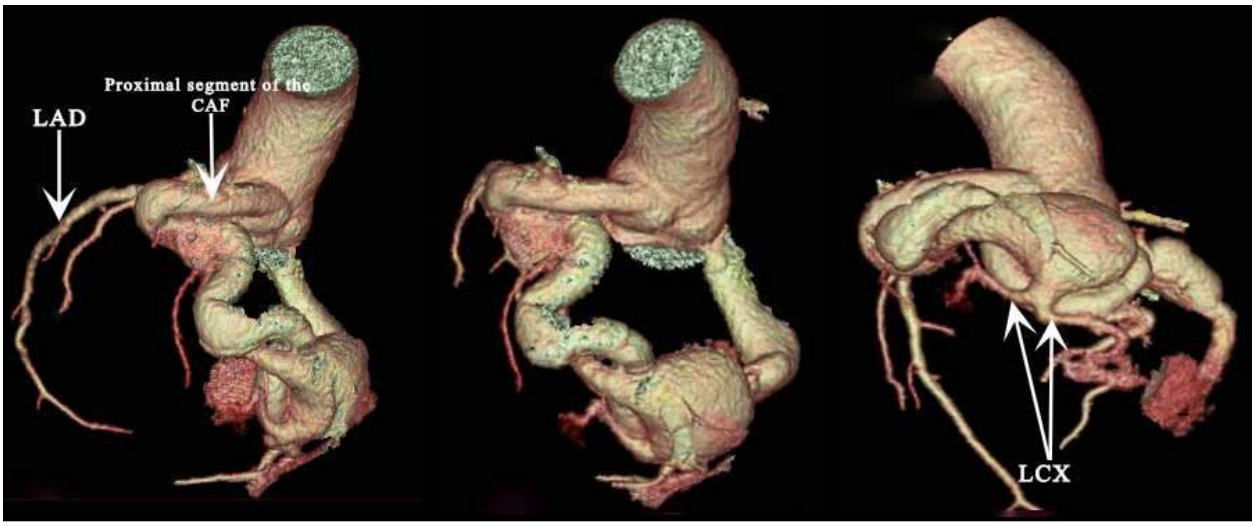


*Figure 1 : 3-D Reconstruction of the CAF*

The CAF Gives LAD as the first branch. Then, the tract having tortuous course in relation to the atrio-ventricular groove. Further, it turns into another aneurysmal segment and gives rise to LCX and its branches. Further, the fistulous tract travels posteriorly and the greater cardiac vein connects into it. Then, the fistulous tract finally drains into right atrium via the dilated coronary sinus. The walls of the fistulous tract showed minor calcific atheromatous plaque disease.

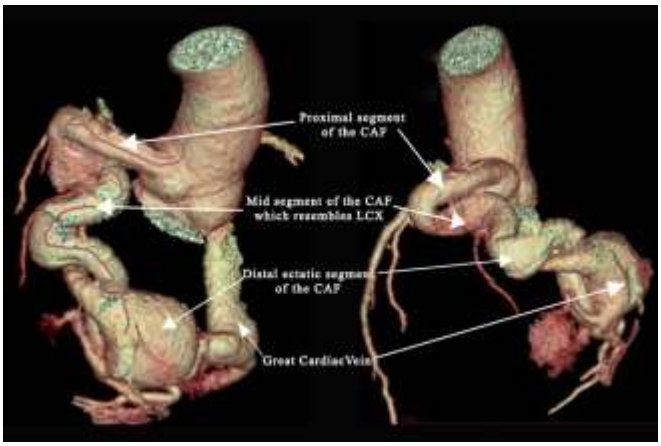


*Figure 2: 3-D Reconstruction of the CAF*

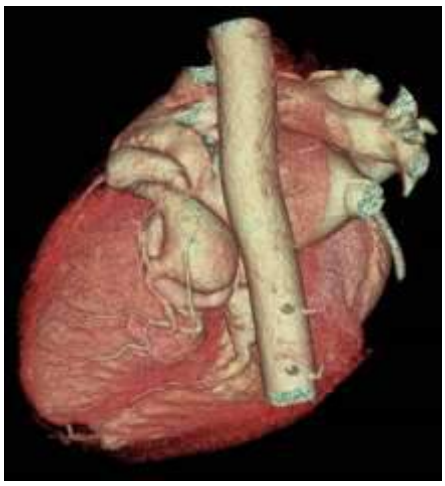

*Figure 3 : 3- D Reconstruction in relation to Aorta*

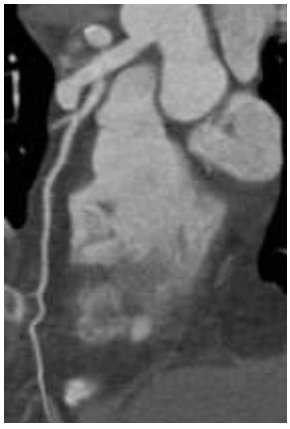

*Figure 4 : LAD with no plaque disease*

As this patient was symptomatic and the fistulae was hemodynamically significant, she was subjected for fistulous tract ligation.

**Case 2**

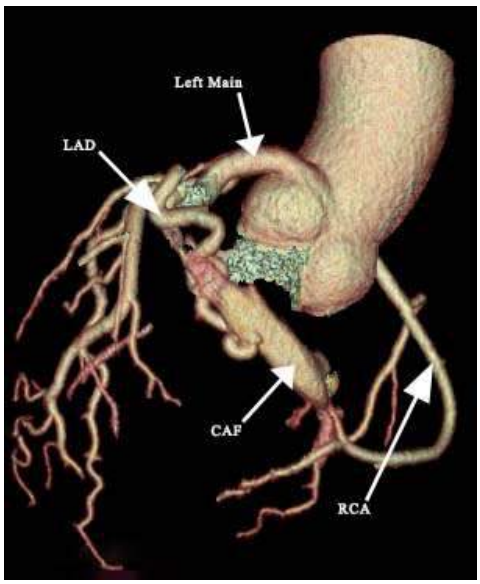


*Figure 5 : 3-D Reconstruction of the CAF - Case 2*

A 42 year old female presented with atypical chest pain for 3 months duration and a physical examination was unremarkable. Her ECG did not reveal any ischemic changes. Her transthoracic echocardiogram was unremarkable except a mild Mitral Regurgitation. A CT Coronary Calcium Score was 0 AU. CT Coronary Angiogram revealed a fistulous tract lies in the posterior atrioventricular groove resembling the course of LCX. The tract is tortuous and large Caliber in nature which has ectatic segments and runs epicardially. The vessel has a fistulous connection into the greater cardiac vein and connects to the enlarged coronary sinus. The middle cardiac vein drains in normal course into the coronary sinus via the greater cardiac vein. The tract does not have atheromatous plaque disease, in-situ thrombi. The fistulous tract finally connects into right atrium via coronary sinus.

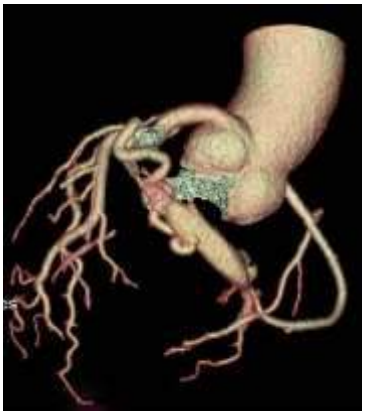

*Figure 6 : 3 D Reconstruction of the Fistulous Tract*

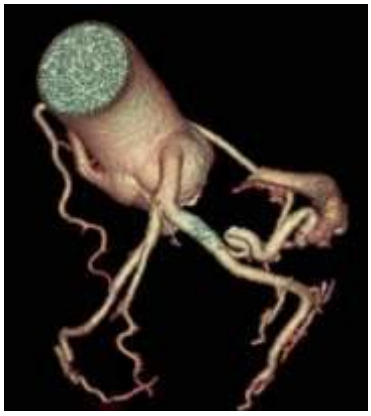

*Figure 7 : 3 D Reconstruction of the fistula*

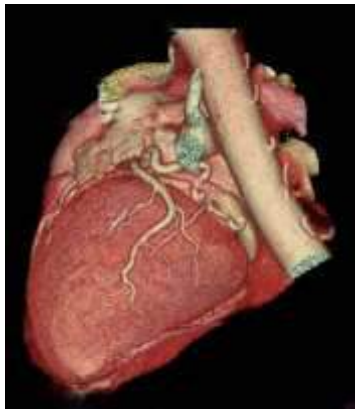

*Figure 8 : 3- D Reconstruction in relation to Aorta*

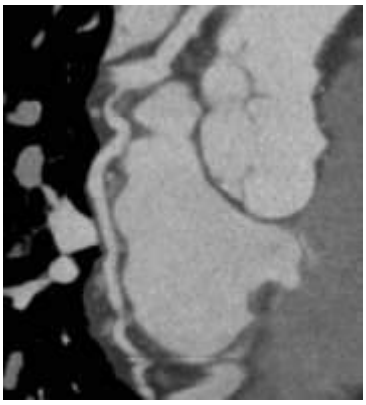

*Figure 9 : Fistulous tract without atherosclerotic plaque disease*

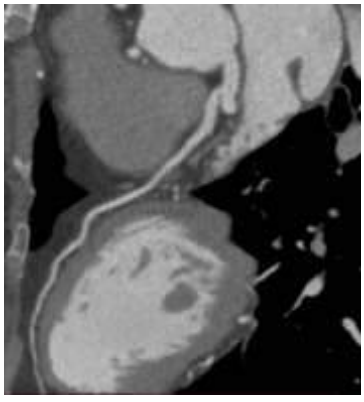

*Figure 10 : LAD without atherosclerotic plaque disease*

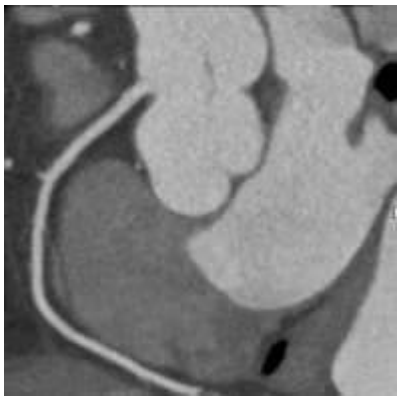

*Figure 11 : RCA without atherosclerotic plaque disease*

As the patient was asymptomatic with a hemodynamically insignificant fistulous tract she was subjected for conservative management with close observation.

**Discussion**

Historical data on prevalence of CAFs from angiographic cohorts is typically about 0.1%. However, contemporary CT-based cohorts report a higher prevalence up to 0.9%. Lim and colleagues found the most common fistula to be coronary artery to pulmonary artery; however in this CT-based cohort, the authors may have been less likely to detect direct connections from coronary artery to a cardiac chambers [1].

Diagnosing modalities of CAFs include Cardiac CT, Cardiac MRI and Invasive Coronary Angiogram. Here, we mainly focus on CT Coronary Angiogram and its characteristics as the main imaging modality.

CT can readily detect surplus vessels but cannot readily detect a direct connection through the myocardium without the advantage of seeing active flow of contrasted blood from the coronary artery into the heart chambers.

In the vascular physiology point of view, the fistulous tract may lead to difference in intra coronary pressure and alters the vascular endothelial physiology. This has the potential result in myocardial ischemia in various aspects. In addition to that these fistulous tracts are comparatively large structures which leads to slow flow and stasis which results in in-situ thrombi formation.

Patients with incidentally found, asymptomatic left-to-left fistulae can be managed conservatively and will often remain stable and symptom-free for long periods of time [1].

If patients present with heart failure, volume overload or ischemia symptoms, the traditional evaluation often includes coronary angiography followed by CTCA, which are the most reliable method of detecting coronary fistulae.

Interventional occlusion or surgical repair is indicated for patients with symptoms attributable to the fistula or heart failure/ cardiac remodeling caused by shunting of blood. Successful repair can usually be achieved with low likelihood of residual shunting in long-term follow-up.

**Conclusion**

CT coronary angiography by itself can provide detailed anatomical delineation of CAFs especially those are having very complex and tortuous anatomy. Therefore, CTCA assists in implementation of critical therapeutic decisions in patients with complex coronary artery fistulae in various clinical setting.

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# A CASE OF NON - COMPACTION CARDIOMYOPATHY: A RARE FORM OF CARDIAC MUSCLE DISORDER

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## Abstract

Left Ventricular Noncompaction Cardiomyopathy (LVNC) is a rare form of congenital cardiomyopathy which is characterized by a thicker myocardium with two layers made up of non-compacted myocardium and a thin layer of compacted myocardium. In this phenomenon, there is an altered myocardial wall architecture with prominent trabeculae and extensive intertrabecular recesses.[6] Heart failure, arrhythmias, which result in sudden cardiac death, and systemic embolic events make up the typical triad of complications. This case study is about a 40-year-old gentleman who presented with heart failure and was subsequently diagnosed to have LVNC.

## Introduction

This is an uncommon cardiomyopathy which is known as left ventricular noncompaction cardiomyopathy (LVNC) comprises of a thin, compacted epicardial layer and a large noncompacted endocardial layer, as well as trabeculations and recesses that link to the left ventricular chamber.[6] It is believed to be brought about by the intrauterine halt of cardiac fiber and meshwork compaction, a crucial phase in myocardium formation. It is mostly associated with congenital heart disease, especially with Ebstein anomaly.[16]And, co-exists with neuromuscular disorders, including Barth syndrome, and muscular dystrophies including Becker muscular dystrophy, Duchenne muscular dystrophy, etc.[14] X-linked and autosomal dominant transmissions are frequently observed in families of LVNC. Patients with isolated LVNC were shown to have mutations in the gene G4.5.[3]A LVNC in long run causes complications such as heart failure, systemic thromboembolism, and arrhythmias which can cause sudden cardiac death. The diagnosis can be made by echocardiogram, Cardiac Computer Tomography(CT), and Cardiac Magnetic Resonant Imaging (CMR).

## Case report

A 40-year-old gentleman presented with gradual onset of dyspnoea for a 1-month duration, he denied angina, palpitation, or syncope and he had a strong family history of congenital heart disease. Physical examination revealed bilateral fine end-inspiratory crepitation in lung bases and elevated jugular venous pressure. Other system examinations were unremarkable. He was hemodynamically stable. Electrocardiography (ECG) found sinus tachycardia with a Left ventricular hypertrophy pattern (Figure 1). His chest X-ray showed cardiomegaly and pulmonary congestion.

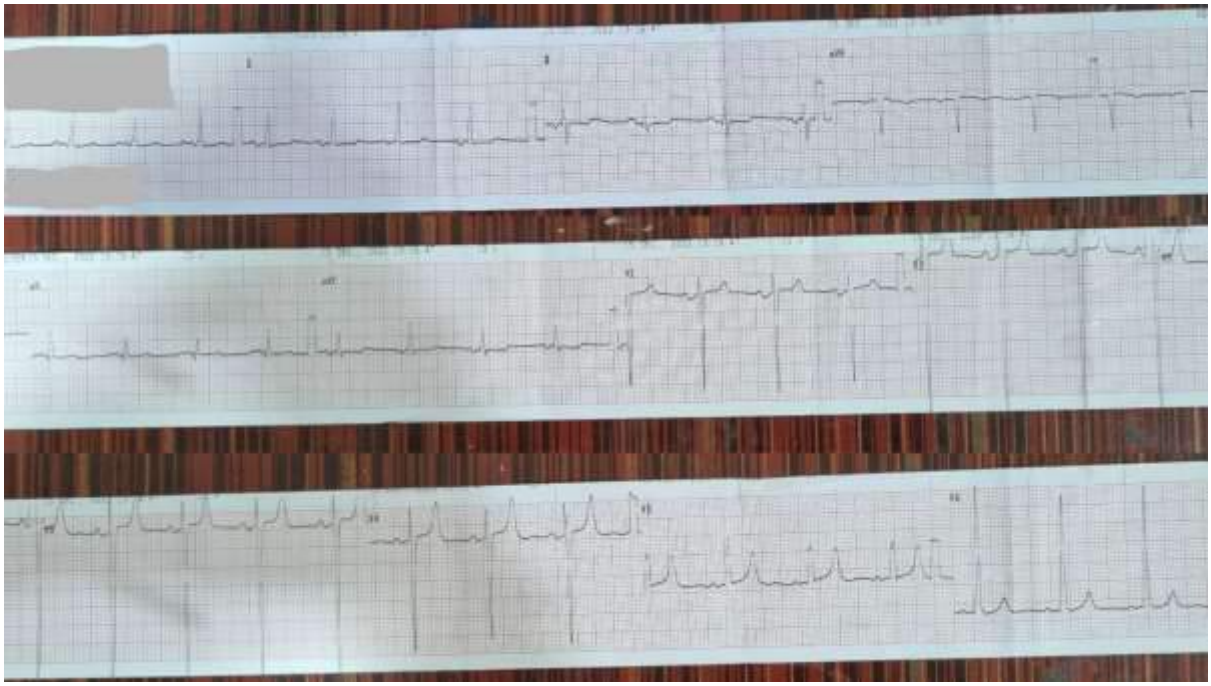


Figure 1: Electrocardiogram showing sinus tachycardia with left ventricular hypertrophy

His echocardiography showed dilated Left Ventricular (LV) cavity with global hypokinesia having an Ejection fraction (EF) of less than 30%. Interestingly, the right ventricle and the left ventricular apex, anterior, and lateral regions involved deep intra-trabecular recesses (Figure 2), which were characterized by many conspicuous trabeculations. And Colour Doppler displayed flow within the deep intertrabecular recesses (Figure 3 and Figure 4). The ratio of non-compacted myocardium to compact myocardium at the end of systole was > 2:1(Figure 5). His Left Atrium appeared dilated with moderate functional Mitral Regurgitation (Figure 6). He also had severe pulmonary Hypertension with Moderate tricuspid regurgitation TRPG- 83 mmHg (Figure 7).

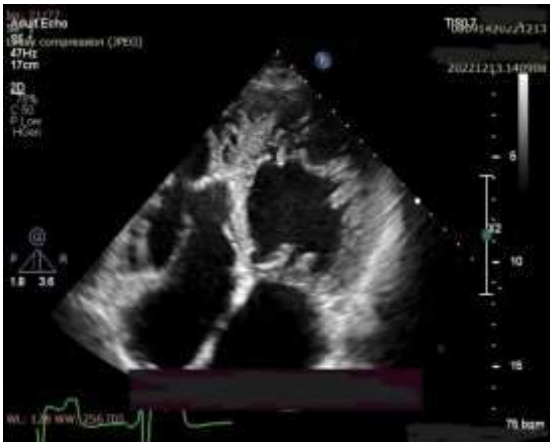


Figure 2: trabeculations involving the left ventricular apical and lateral wall and right ventricle

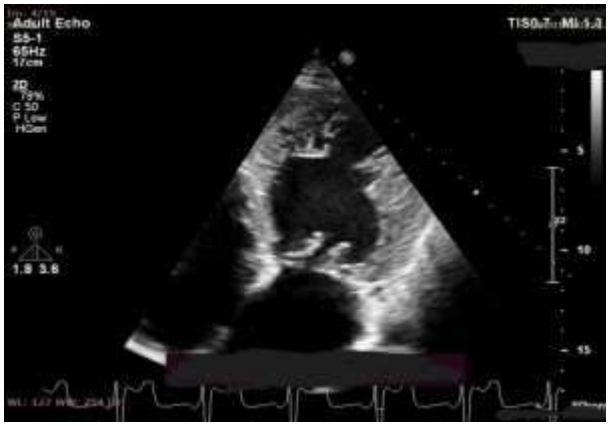



Figure 3: Left ventricle with trabeculations

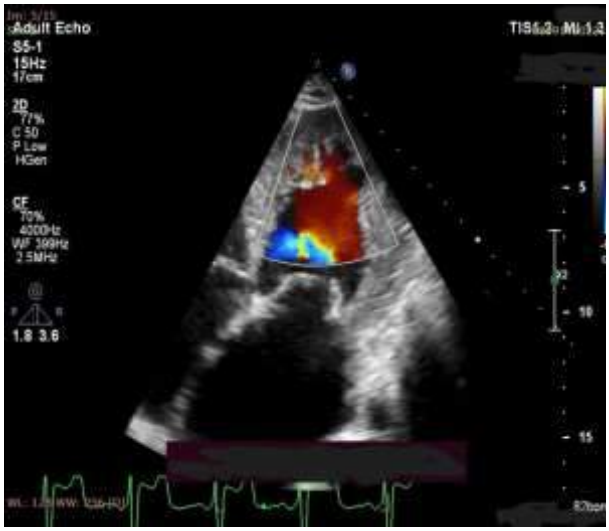



Figure 4: colour doppler flow within the deep intertrabecular recesses

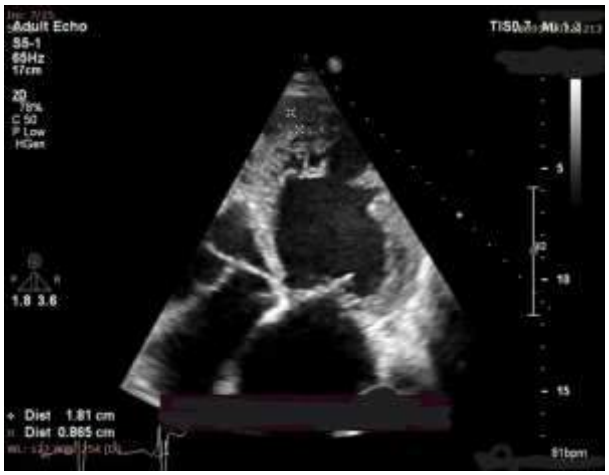
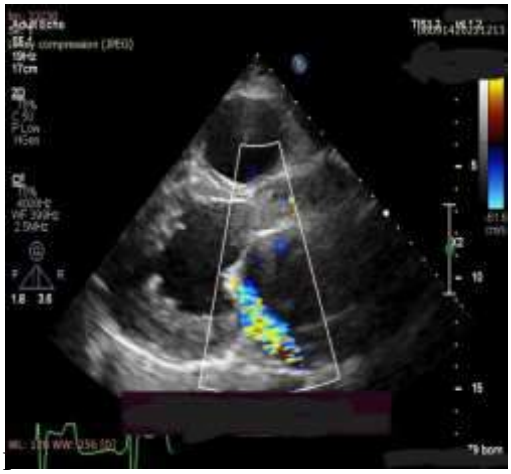



Figure 5: The ratio of noncompacted myocardium to compact myocardium at the end of systole > 2:1



Fiigure 6: Left atrium dilated with moderate functional Mitral regurgitation

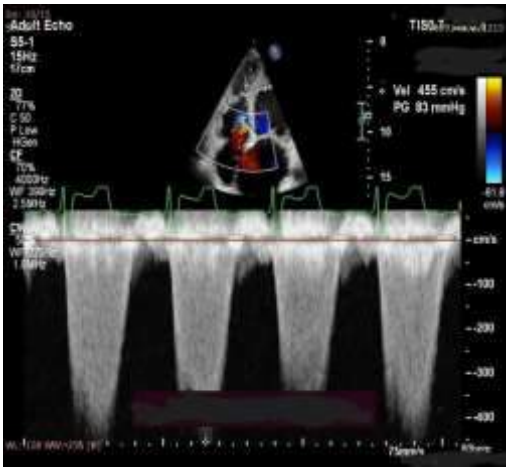



Figure 7: Moderate tricuspid regurgitation wi th Severe pulmonary hypertension

He was started with an anti-failure regime for offloading. After the optimization of medical management, his symptoms settled with the improvement of his functional capacity. The Coronary angiogram revealed normal epicardial coronaries and the 24-hour Holter study showed no significant arrhythmia. Once he is off symptoms, warfarin was commenced in view of prevention of potential thromboembolism associated with abnormal hemodynamics in LVNC in the background of severe LV systolic dysfunction.

**Discussion**

Due to its spongy appearance, LVNC was formerly referred to as "Spongiform Cardiomyopathy." This is rather rare entity and LVNC was found in 0.014-1.3% of patients undergoing echocardiography. [15]

There is an echocardiographic criterion to diagnose LVNC. According to a study of 34 post-mortem specimens by Jenni et al, the following criteria for diagnosing LVNC were developed. (1) Absence of coexisting cardiac abnormalities (2) A thin, compacted epicardial band and a much thicker, non-compacted endocardial layer of trabecular meshwork with deep endomyocardial gaps. It is diagnostic to have a maximum end-systolic ratio of non-compacted to compacted layers of > 2. (3) Apical, mid-lateral, and mid-inferior regions were the most affected by the pathology. (4) Deep perfused intertrabecular recesses were visible using the colour Doppler.[5]The presence of all four of the echocardiographic criteria are required for the diagnosis of LVNC. This patient fulfilled all these criteria in his echocardiogram.

CMR can reliably identify LVNC by measuring the Noncompaction (NC) / Compaction (C) ratio in diastole. It is advised to take three diastolic long-axis images in order to diagnose LVNC on CMR and to select the myocardial segment with the most noticeable myocardial trabeculations in order to calculate the NC/C ratio. The diagnosis of LVNC may be supported by CMR black blood imaging, which can find increased signal intensity within the myocardial trabeculae, a marker of stagnant blood flow. According to the study of Petersen et al, the CMR criteria to diagnose (LVNC) was proposed as follows, The inner noncompacted layer displaying two separate myocardial layers, prominent trabeculations, and deep intertrabecular recesses; a NC/C myocardium ratio > 2.3 at end-diastole was deemed suggestive of LVNC. However, it had 86% sensitivity & 99% specificity in the diagnosis of LVNC.[8]

There are some additional CMR criteria to diagnose LVNC as well. As Jacquier et al recommend for the diagnosis of LVNC as trabeculated left ventricular mass should be more than 20% of the global left ventricular mass measured at end-diastole.[4] A quantitative assessment for the diagnosis of LVNC was developed by Grothoff et al. which is Trabeculated mass should be greater than 25% of the LV's overall mass and greater than 15 g/m2.[2]

For the detection of non-compacted myocardium, CMR imaging is superior to echocardiography due to greater image quality and increased sensitivity for detecting trabeculations, especially during end-diastole.[1] It was established that CMR plays a key part in the evaluation of these patients when the diagnosis by the echocardiography is not confirmed due to a lack of image quality. Additionally, the degree of fibrosis may help in distinguishing the severity of the disease in CMR. In our case, CMR is not available at our facility, nevertheless, our

echocardiogram provided good-quality images to diagnose LVNC. A CMR study found that LVNC is associated with increased trabeculations of the RV apex and that RV dysfunction in an LVNC population is associated with adverse clinical events.[11]

Cardiac Computer Tomography is not an excellent test to diagnose LVNC. Even though it is used in some setups where the echocardiography is indeterminate or technically suboptimal and when CMR is unavailable. An end-diastolic noncompacted to the compacted ratio of >2.3 on CT long-axis views were found to be the best marker for LVNC.[10] Although there are no well-proposed criteria to diagnose LVNC by Cardiac Computed Tomography.

LVNC gives rise to more devastating complications in the long term such as heart failure, ventricular tachycardia, thromboembolic events, and sudden cardiac death. [7] Our patient managed with an anti-failure regime to control heart failure and the Holter study does not show significant arrhythmia. Moreover, warfarin was commenced to avoid thromboembolism. In advanced settings, some patients undergo heart transplantation and implantation of an automated defibrillator/cardioverter which enhances the prognosis.

Importantly, the overall mortality did not differ significantly between patients with isolated LVNC and control patients with dilated cardiomyopathy (3-year survival of 85 vs. 83%). [12]

**Conclusion**

Although LVNC is a rare form of cardiomyopathy, it's important to consider it as a differential diagnosis when dealing with dilated cardiomyopathy. The Hallmark of diagnosis is the presence of trabeculations. And when cardiomyopathy is associated with congenital heart disease and neuromuscular disorder, it's very crucial to consider LVNC as a differential diagnosis. Moreover, the management of LVNC includes a heart failure treatment, identifying arrhythmic conditions, and preventing thromboembolic phenomena. The recent advancement of technology aids in the diagnosis of LVNC. In the past, the diagnosis mostly depended on echocardiography but nowadays it is diagnosed more specifically by CMR.

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# RARE CASE OF THYROTROPH HYPERPLASIA SECONDARY TO PRIMARY HYPOTHYROIDISM MIMICKING A PITUITARY ADENOMA: A CASE REPORT AND A REVIEW OF LITERATURE

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**Abstract**
Primary hypothyroidism can lead to uncommon presentations like tumorous signs secondary to pituitary thyrotroph hyperplasia. Imaging studies may not be able to differentiate hyperplasia from pituitary adenoma. We present a case of A 9-year-old child who presented with headache and vomiting, found to have a pituitary mass on the MRI with high TSH values. After thyroxine replacement and normalization of TSH, pituitary mass resolved on follow-up MRI, confirming pituitary hyperplasia. Thyrotroph hyperplasia may mimic pituitary adenoma but careful biochemical analysis would indicate the underlying primary hypothyroidism. Appropriate treatment with thyroxin may resolve pituitary hyperplasia which would prevent an unnecessary pituitary surgery.

**Key words** – Pituitary hyperplasia, Primary hypothyroidism, Thyrotroph

## Introduction

Long standing untreated primary hypothyroidism in childhood may present with cretinism, pubertal delays and cognitive effects. Loss of feedback inhibition for thyrotroph releasing hormone (TRH) at hypothalamus in primary hypothyroidism will increase thyroid stimulating hormone (TSH) release from the pituitary. TRH also exhibits trophic effect on pituitary thyrotrophs (1).

Enlargement of the pituitary gland in children occur due to an adenoma of the pituitary gland or pituitary hyperplasia, especially during puberty (2). Pituitary hyperplasia following stimulation by TRH is rare and may occur in long standing untreated primary hypothyroidism (2-5). Patient may present with features of pituitary adenoma, hyperprolactinemia, features of pituitary hormone deficit or problems in growth and puberty (4, 5). Imaging studies alone would not be able to differentiate a TSH producing pituitary adenoma or hyperplasia. Repeat pituitary imaging following treatment with Levothyroxine will reveal resolution of hyperplasia following treatment will confirm the diagnosis and avoid unnecessary pituitary surgery.

We report a case of pituitary enlargement due to an undiagnosed primary hypothyroidism, with subsequent resolution with adequate replacement of thyroxine.

## Case report

A 9-year-old previously healthy boy with uneventful perinatal period and childhood presented to the pediatric clinic with generalized headache for 4 months duration. It was associated with vomiting and dizziness but had no visual impairment. He did not have any seizures, hearing impairment, or focal neurological symptoms. Examination revealed GCS of 15/15, blood pressure 95/65($50^{th}$ -$75^{th}$ Centile) and age-appropriate weight but a height at $3^{rd}$ centile. He was in pubertal stage I. There were no focal neurological signs and his visual fields, pupils and optic fundi were normal. Basic laboratory investigations (Full blood count, C-reactive proteins, erythrocyte

sedimentation rate, serum electrolytes) were normal.

Magnetic resolution imaging (MRI) brain showed a pituitary macroadenoma (1.5 x 1.4 x 1 cm) arising from the adeno hypophysis with suprasellar extension causing mild pressure effects on the post optic nerve and chiasma (Figure 1). Following the detection of the pituitary lesion, hormone panel was performed, which revealed markedly elevated TSH and raised prolactin. (Table 1).
Differential diagnoses at this point were TSH producing pituitary macroadenoma or Pituitary macroadenoma with primary hypothyroidism. As the TSH levels were markedly elevated and there were no clinical features of visual field defect, Levothyroxine was initiated as for primary hypothyroidism with careful TSH monitoring and thyroxine dose adjustments.

Table 1: Pituitary hormone level change with time

| | 2017 Aug | 2018 Feb | 2022 Dec |
|---|---|---|---|
| 9am cortisol [nmol/L] | 403 (138-635) | - | - |
| LH [µIU/ml] | <0.2 (0.3-6) | - | 1.6 |
| FSH [µIU/ml] | 2.0 (1-5) | 0.07 | 3.4 |
| Prolactin [ng/dL] | 85 (0-20) | 12 | 14 |
| IGF-1 [ng/ml] | 124.2 (74-388) | - | - |
| TSH [µIU/ml] | >100 (0.4-4) | 5.68 | 3.2 |

Repeat MRI brain February 2018 showed interval reduction of the pituitary macroadenoma compared to the previous study. Further evaluation by a follow-up MRI brain in February 2021 showed a normal pituitary gland (0.6 x 0.5 x 0.8mm) without any tumor or enlargement (Figure 2)

With time, his height also has improved (from 25$^{th}$ centile to 75-95$^{th}$ centile) together with his development of secondary sexual characteristics.

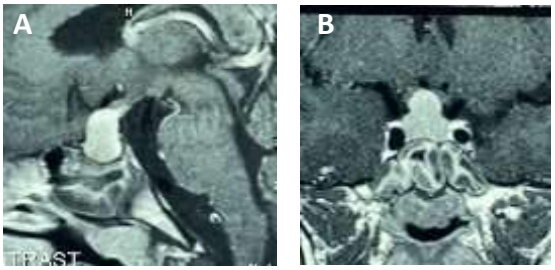


Figure 1- A – Sagittal view of Pituitary hyperplasia mimicking a pituitary adenoma. B – Coronal view of pituitary hyperplasia mimicking a pituitary adenoma

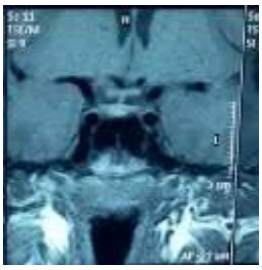

Figure 2 - Normal pituitary gland following treatment with levothyroxine and normalization of TSH

**Discussion**

Longstanding primary hypothyroidism leads to pituitary hyperplasia caused by loss of negative feedback from low circulating thyroxine (T4) and triiodothyronine (T3) hormone levels, leading to excess TRH which intern stimulates pituitary thyrotrophs to proliferate as well as to secret TSH. This leads to Pituitary "tumorous" hyperplasia, accounting for approximately 33.3% of cases of secondary pituitary hyperplasia from end organ insufficiencies (6).

Index patient presented with tumoral signs and symptoms and this is reported in 25% of patients with pituitary hyperplasia following primary hypothyroidism (7). Among this group, only 38% present with main hypothyroid symptoms and our

patient also did not exhibit classic hypothyroid symptoms apart from reduced height (7). Degree of TSH elevation and level of pituitary hyperplasia has shown a correlation where 84% of patients with TSH >100 µIU/L had shown pituitary hyperplasia like in the index case (8).

Hyperprolactinemia also is a known feature of severe primary hypothyroidism, seen in about 39% of cases (9), which was again observed in our patient. It is due to the stalk effect by pituitary hyperplasia and effective treatment of hypothyroidism will normalize prolactin levels.

Features in MRI alone may not be able to differentiate pituitary hyperplasia vs pituitary adenoma. Only 0.5% of pituitary tumors are TSH secreting, which is very rare and a high level of TSH in patients like in the index case should alarm the physicians about possibility of pituitary hyperplasia (7). Adequate treatment with thyroxin regresses the pituitary hyperplasia in 85% of patients according to the literature and can occur as early as 1 week to 1-3 months (8, 10).

## Conclusion

Pituitary hyperplasia in primary hypothyroidism is not uncommon and pituitary enlargement can be frequently observed on imaging. Correct recognition of severe primary hypothyroidism causing pituitary hyperplasia is important to prevent any unnecessary surgical intervention. Both radiographic imaging results and biochemical testing results are essential prior to decision of treatment modality. Patients with hypothyroidism, who have pituitary enlargement diagnosed on brain imaging, should be treated with thyroid hormone replacement and close follow-up with repeat imaging and hormonal profile is warranted.

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# HOW DOES CARDIAC IMAGING GUIDE TO DELINEATE THE CORONARIES WITH ANOMALOUS ORIGIN?: A CASE SERIES

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**Abstract**

Anomalous Coronary Arteries are rare forms of congenital anomalies. Of these, Right Coronary Artery (RCA) arising from the left coronary cusp has been said to be commoner than Left Coronary Artery (LCA) arising from right coronary cusp while single common ostium for all three coronaries has been found to be extremely rare. Anomalous Coronary Artery arising from the Pulmonary Artery (ALCAPA) accounts for about 0.25-0.5% of congenital cardiac diseases. Majority of these are asymptomatic, but some may present with symptoms due to arrhythmias or myocardial ischemia or even with complications such as myocardial infarction or sudden cardiac death.

Diagnosis of these mainly depend on cardiac imaging modalities. Computed Tomography Coronary Angiography (CT-CA) is an excellent method in identifying and describing these anomalies. 2D Echocardiography is ideal for children as it prevents exposure to harmful radiation. Magnetic Resonant Angiography (MRA) facilitates evaluation of the extent of ischemia and the viability of the heart muscle but is less useful in assessment of coronary vessels. Invasive Coronary Angiography (ICA) can mainly diagnose the atherosclerotic burden of the coronaries along with their anomalous course. Each of these have their own strengths and weaknesses which guide us on analysis of the anatomy and decision making with regard to further management and follow-up.

We present a series of four cases of patients having coronaries with anomalous origins and how the management depended on the symptom profile and the anatomy described by the relevant imaging modalities.

## Introduction

The coronary circulation originates from the sinuses at the beginning of the aortic root where the Right Coronary Artery (RCA) arises from the right anterior sinus and a Left Main Coronary Artery (LMA) arising from the left anterior sinus and further divides into Left Circumflex Artery (LCX) and Left Anterior Descending Artery (LAD).

Congenital Coronary artery abnormalities are a broad category of diseases with a wide range of pathophysiological causes and clinical presentations. These are mainly categorized into four, depending on the origin and course, termination, intrinsic anatomy and anomalous anastomotic vessels. [1]

Many patients are asymptomatic and these anatomical variations are incidental findings. However, one should be cautious in patients presenting with anginal symptoms on exertion which cannot be explained by other cardiovascular risk factors, especially in the young. Symptomatic individuals may be subjected to a range of treatment modalities from observation with medical management to coronary angioplasty with stenting or surgical repair.

The definition, morphogenesis, clinical presentation, diagnosis, treatment and prognosis of coronary artery anomalies are still undergoing a significant evolutionary change. [2] A prospective analysis done by studying coronary angiograms of 1950 patients showed a global incidence of 5.64% in coronary artery anomalies. Of them, total incidence of Anomalous origination of coronary artery from the opposite sinus had a total incidence of 1.07%, where anomalous origin of RCA from Left sinus had an incidence of 0.92% and anomalous origin of LCA from right sinus of 0.15%. [3] In a study done using angiography results of 126,595 patients, majority of anomalous origins were of RCA (0.17%) than of the LCA (0.047%). [4]

The latter has been revealed to be accounted for up to 85% of sudden cardiac deaths associated with anomalous coronary artery origins. [5]

Current proposed theories on deaths due to coronary artery anomalies are ischemia induced by kinking of coronary arteries due to the acute angulation at the ostium, narrow opening, malignant course between the aorta and pulmonary artery leading to a mechanical compression or due to vasospasms. [6]

Transthoracic or transesophageal Echocardiography are few of the basic investigations which play a major role in initial diagnosis. In conditions such as Anomalous origin of LCA from Pulmonary Artery (ALCAPA), 2D echocardiography findings strengthens its diagnosis preventing the risk of exposure to radiation as well. However, more sophisticated, invasive investigations such as Coronary Angiography, Cardiac Computed Tomography or Cardiac Magnetic Resonance Imaging help in better elaboration of these anomalies. [3], [7], [8]

Currently, an ideal imaging tool for the diagnosis of the coronary artery anomalies is Computed Tomography Coronary Angiography (CT-CA) which is a non-invasive procedure which gives rise to highly descriptive images. [9]

In certain instances, intervention may be appropriate to avert unexpected death and enhance quality of life despite some limitations. Medical treatment such as beta blockers, close observation with restriction of activity, stent deployment following coronary angioplasty (depending on the degree of obstruction/stenosis of the vessels by atherosclerosis) or surgical repair are the main treatment modalities. [10]

We describe a series of cases which highlights the importance of 2D echocardiography and CT-CA which guided in the delineation of anomalous origin of coronary arteries.

**Case 1 – (Malignant course of RCA)**

A 47 years old previously healthy male presented with multiple events of recurrent atypical type chest pains. Physical examination was unremarkable. ECG did not depict any ischemic changes and 2D echocardiogram showed a structurally normal heart. The Exercise Tolerance Testing (ETT) was terminated prematurely at stage II due to a mechanical type leg pain. Therefore, he was subjected to a CT-

CA, as he had overall low probability to have coronary artery disease.

His CT-CA showed Anomalous origin of a non-dominant RCA from left coronary sinus with subsequent course between the aorta and the pulmonary trunk; the malignant course of RCA, with no intramural segment or kinking at the RCA ostium. The dominant LCX and LAD had normal origin and course. None of the coronaries had atheromatous plaque disease.

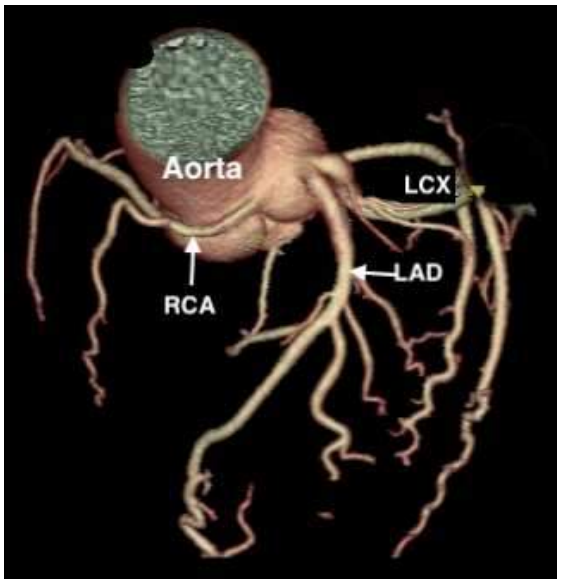



**Figure 1.1: Right Coronary Artery (RCA) arising from the Left coronary sinus of the Aorta. LAD: Left Anterior Descending Artery, LCX: Left Circumflex Artery**

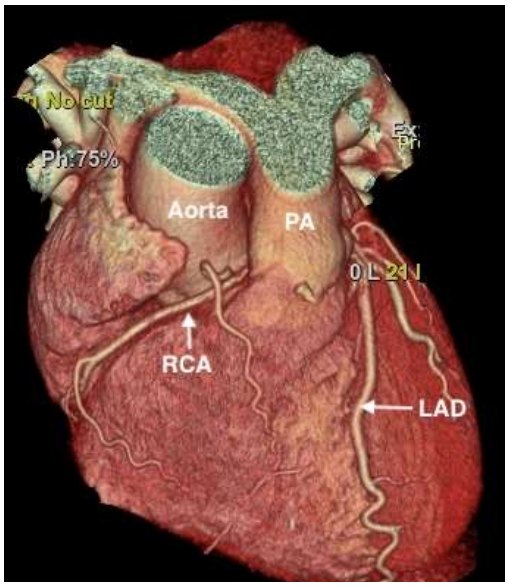



**Figure 1.2: Course of Right Coronary Artery (RCA) in between Aorta and Pulmonary Artery (PA). LAD: Left Anterior Descending Artery**

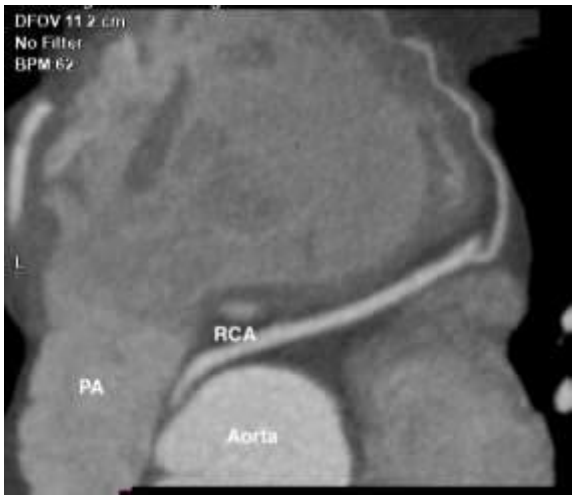



**Figure 1.3: Right Coronary Artery (RCA) not showing any evidence of atherosclerotic plaque disease. PA: Pulmonary Artery**

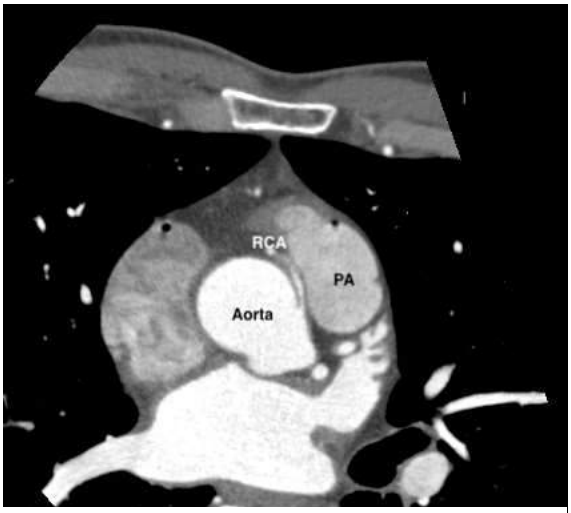



**Figure 1.4: Right Coronary Artery (RCA) arising from the Aorta and lying in-between Aorta and Pulmonary Artery (PA)**

Subsequently he was subjected to Dobutamine Stress Echocardiogram (DSE) as a functional test and it was negative. He was conservatively managed with close observation because even though his RCA had a malignant course, but had no evidence of inducible ischemia or other high-risk features in the CT-CA.

## Discussion

Incidence of anomalous origin of the RCA, from the left coronary cusp with a malignant course varies between 0.026% and 0.250%. This course between pulmonary artery and aorta lead to a risk of RCA being compressed during strenuous or sometimes routine activities, which may lead to angina, arrhythmias leading to syncope, or even sudden cardiac deaths. [11]

In addition to this usual malignant form, other known features which increase the risk are the intramural course where the proximal vessel passes through the tunica media of the aortic wall with narrowing, the acute angle at the origin and the slit-like ostium. [12]

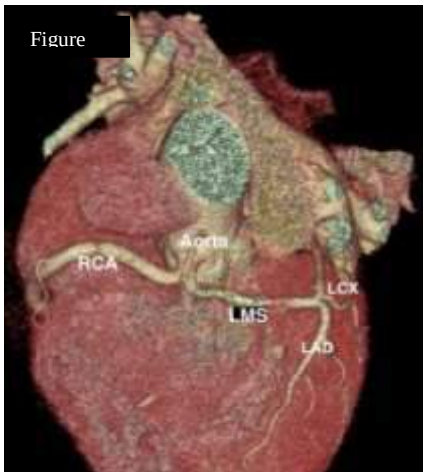
Diagnostic tools of choice are said to be CT-CA and MRI Coronary Angiography, where CT-CA provides higher quality images with more details including coronary artery disease. [13] Prevalence of these anomalies depends on the diagnostic modality used; 0.15% in echocardiogram, 0.70% in MRA and relatively highest in CT angiography which is 0.82-1%, concluding CT-CA as an important modality in diagnosis. [14]

Indications for surgical interventions mainly depend on factors such as ischaemic symptoms and the evidence of high-risk features in coronary imaging. Surgery is performed to correct the intramural route and accompanying ostial stenosis by un-roofing, ostioplasty, coronary artery bypass grafting or translocation and reimplantation. [15]

## Case 2 (LCA originating from RCA - common origin)

A 41 year old previously healthy female presented with recurrent episodes of non-specific chest pain. She had no other vascular risk factors other than obesity. Examination was unremarkable. ECG did not depict any ischemic changes and 2D echocardiography revealed a structurally normal heart. A CT-CA was performed to assess her coronaries as her overall probability to have Coronary Artery Disease (CAD) was low.

Her CT-CA showed a single common origin of all three coronary arteries from the right coronary cusp through a common trunk. Her RCA was in normal course and Left Main Stem (LMS) originated from RCA. Proximal segment of LMS lied posterior to pulmonary trunk where it divided into LAD and LCX. There was no evidence of flow limiting coronary plaque disease in any of the vessels.



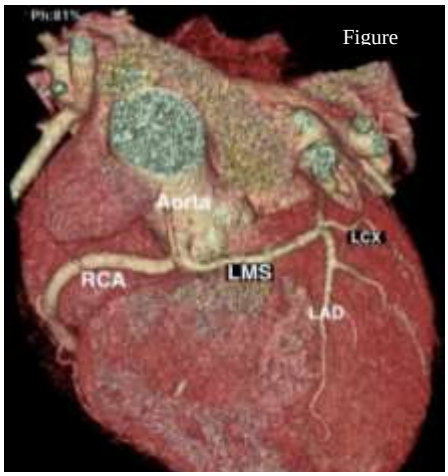
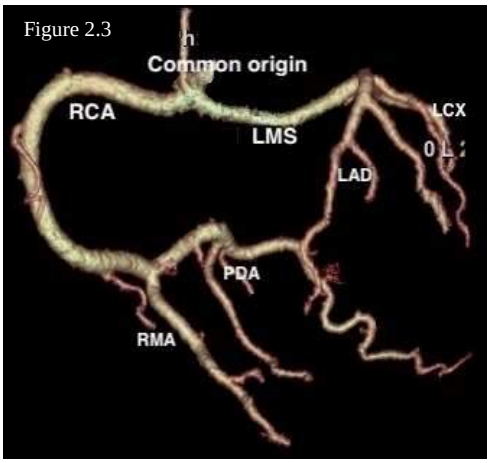




**Figure 2.1 and Figure 2.2: Right Coronary Artery (RCA) and Left Main Stem (LMS) arising as a common origin from the Aorta. LAD: Left Anterior Descending Artery, LCX: Left Circumflex Artery**



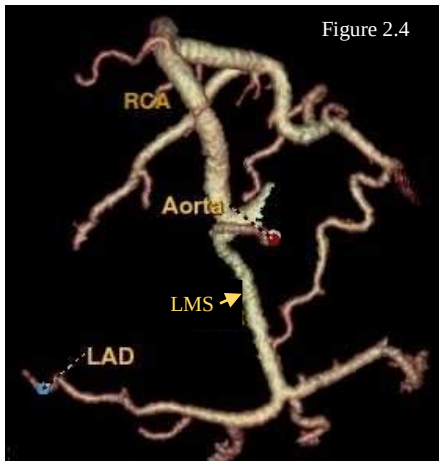




**Figure 2.3 and Figure 2.4: All coronary arteries arising from a common origin, Right Coronary Artery (RCA) in its normal course divides in to Right Marginal Artery (RMA) and Proximal Descending Artery (PDA), Left Main Stem (LMS) originating from the RCA and dividing into Left Anterior Descending Artery (LAD) and Left Circumflex Artery (LCX)**

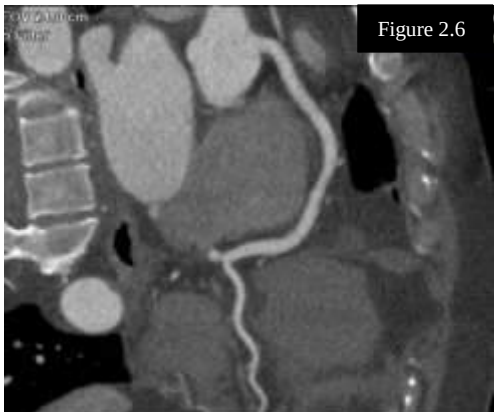




**Figure 2.5 and Figure 2.6: Coronaries not showing any evidence of flow limiting atherosclerotic plaque disease**

She underwent ETT for further evaluation and was able to complete stage IV without any evidence of inducible ischemia. Therefore, it was decided to keep her under conservative approach.

## Discussion

Various Anomalies in coronary arteries, being rare congenital disorders, a single common ostium for all three coronary arteries is a highly uncommon variation, with a prevalence of under 0.004%.[16] Patients may be asymptomatic or have ischemic symptoms and higher risk of sudden cardiac deaths. It's said that atherosclerosis is common in coronary arteries which have an anomalous origin and course. However, luckily this patient did not have atherosclerosis in the common trunk as it would be difficult to intervene in view of Percutaneous Coronary Intervention (PCI). Our patient presented at the age of 41 years whereas literature indicates 35 years as the average age at which patients present with symptoms and the oldest alive was found to be 83 years. [17]

This scenario also depicts the importance of CT-CA which serve many benefits including avoidance of conventional invasive methods of coronary angiography while facilitating the assessment of functional and hemodynamic significance

of these anomalies along with long term follow up. [18]

Mainstay of treatment for high risk patients with indications is surgical management in view of preventing myocardial ischemia. Major surgical interventions include, un-roofing of vessels with extensive intramural course and ostioplasty or ostial translocation (re-implantation) if only a short intramural course is present. Coronary artery bypass grafting is suggested if concomitant atherosclerotic narrowing coexists or if other interventions have failed. [19]

**Case 3 (Echocardiographic diagnosis of ALCAPA)**

This is a case of a 17 year old girl, who's antenatal period was uneventful. At 23 days of life she developed a breathing difficulty along with heart failure features. Her subsequent 2D echocardiograms revealed a dilated Left Atrium (LA) and Left Ventricle (LV), severe Mitral Regurgitation (MR) with Ostium Secundum Atrial Septal Defect (OS-ASD) with left to right shunt. She was started on anti-failure regimen. She was planned for a Mitral Valve repair but during the serial echoes, at the age of 7 years, she was found to have an Anomalous origin of Left Coronary Artery from the Pulmonary Artery (ALCAPA), with preserved cardiac functions due to multiple collaterals.

Currently she is awaiting corrective surgery. Despite these echo findings, she remains active and asymptomatic. Her examination findings remain unremarkable except for the Grade II MR murmur heard at the apex. ECG showed sinus rhythm with no abnormal ST, T or Q wave changes.

In 2D Echocardiogram, the ostium of LCA at the Aortic root was absent, depicting The LCA arising from Main pulmonary trunk towards the aortic root. RCA was dilated. Increased vasculature in the Inter-Ventricular Septum (IVS) noted which are multiple collaterals from RCA to LCA, and this mimics multiple Ventricular Septal Defects (VSD). Apart from these findings, she also had a Grade II MR with Mitral Valve Prolapse (MVP) and marginally dilated LA and LV.

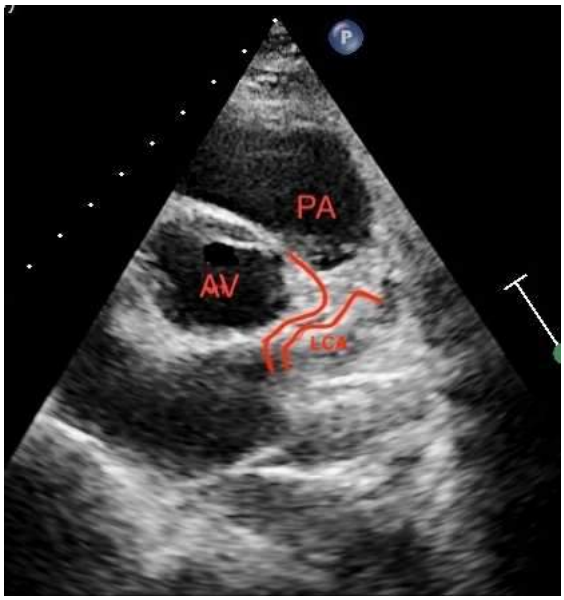



**Figure 3.1: Anomalous origin of the Left Coronary Artery (LCA) from the Pulmonary Artery (PA) (marked by the red line). Ostium of LCA is not seen at the Aortic root. AV: Aortic Valve**

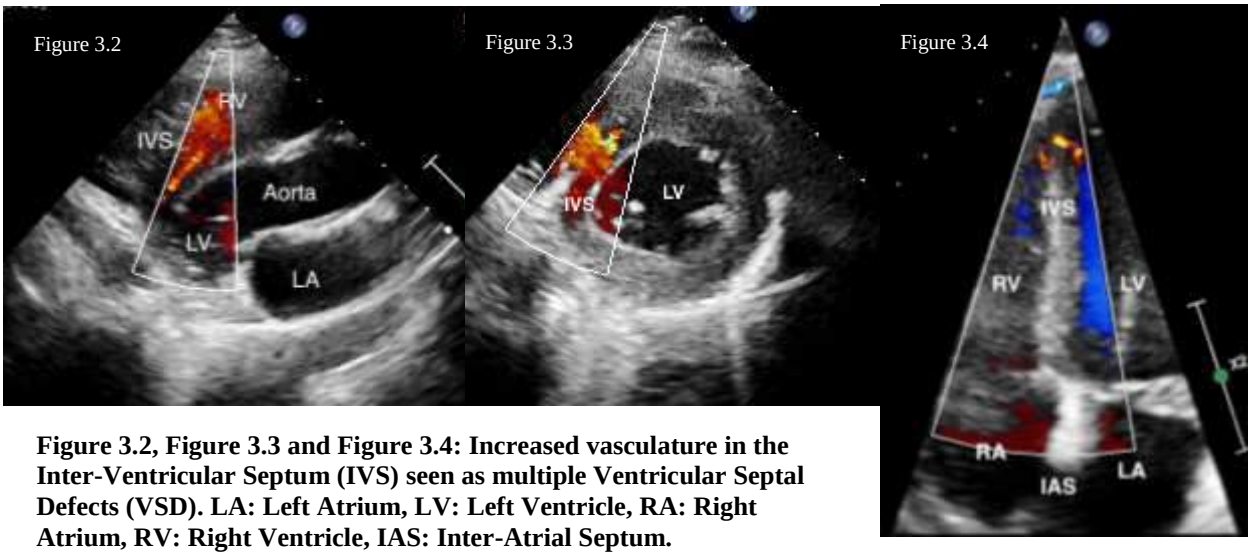



**Figure 3.2, Figure 3.3 and Figure 3.4: Increased vasculature in the Inter-Ventricular Septum (IVS) seen as multiple Ventricular Septal Defects (VSD). LA: Left Atrium, LV: Left Ventricle, RA: Right Atrium, RV: Right Ventricle, IAS: Inter-Atrial Septum.**

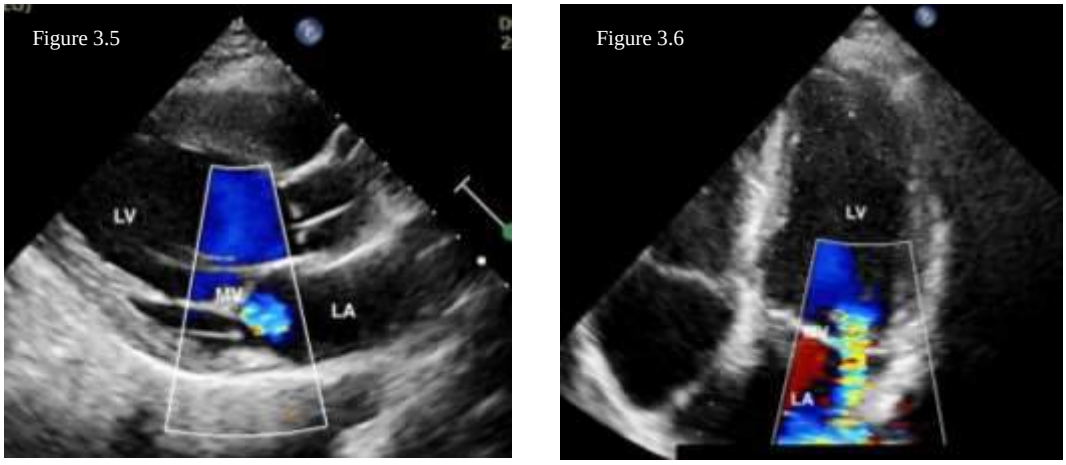



**Figure 3.5 and Figure 3.6: Grade II Mitral Regurgitation. LA: Left Atrium, LV: Left Ventricle, MV: Mitral Valve**

## Discussion

ALCAPA is a rare congenital anomaly which affects 1 in 300 000 live births and accounts for 0.25 to 0.5% of all congenital cardiac diseases. Anomalous origin of the Right Coronary Artery from the Pulmonary Artery (ARCAPA), is a very rare anomaly which accounts for about 0.002% of all congenital cardiac diseases. [20]

Usually it is diagnosed during the childhood, predominantly in infancy, presenting with features of congestive heart failure and or myocardial ischemia. It is important to differentiate this from diseases with similar presentations such as bronchiolitis, infantile colic or gastroesophageal reflux disease, as good prognosis depends on early diagnosis and intervention. [21] It has a mortality of 90% during the first year of life. [22]

Some survive longer, with or without symptoms up to adulthood, depending on multiple factors such as RCA dominant coronary artery system and development of collaterals from RCA to LCA. Even though asymptomatic, underlying chronic

subclinical ischemia might be present, leading to MR as in this patient, left ventricular dysfunction or sudden cardiac death. [23]

High pulmonary artery pressures during fetal life cause an antegrade flow in the LCA, which reverses after birth due to reduction of pulmonary vascular resistance. This causes de-saturated blood to enter LCA, leading to myocardial ischemia mainly in the sub-endocardial region.[24] ECG commonly shows inverted T waves in leads I and aVL and deep Q waves, elevated ST segments and inverted T waves in leads V5 to V6. [21] These findings were absent in our patient. Echocardiography features in diagnosis of ALCAPA are absent LMA ostium at the aortic root, shunting flow from LCA ostium to pulmonary artery and vascular collaterals within the inter ventricular septum and free ventricular wall, which is a more prominent feature as in our patient.

In a study done using 30 patients who were already established to be having ALCAPA, 24 of them were diagnosed using echocardiography. The study concluded that echo plays a major role in detecting this coronary anomaly. [25]

**Case 4 (CT Coronary Angiographic diagnosis of ALCAPA)**

A 48 year old lady who presented with exertional dyspnea was found to have a positive ETT. Her 2D echocardiography showed a Dilated LA and LV with preserved Ejection Fraction (EF). As her ETT was positive, she had undergone an ICA which showed RCA with septal collaterals. The LAD ostium was not visualized.

Her CT-CA showed RCA originating from Right coronary cusp and was tortuous and large caliber, giving rise to multiple septal feeding vessels to LAD. The conus artery had almost separate origin from RCA and it also gave multiple septal feeding vessels to LAD. LMA originated from the proximal segment of Pulmonary Artery (PA) which further divided into large caliber LAD which was in the anterior interventricular groove and a moderate caliber LCX which was lying in the atrioventricular groove. None of the vessels had atherosclerotic plaque disease.

Figure 4.1

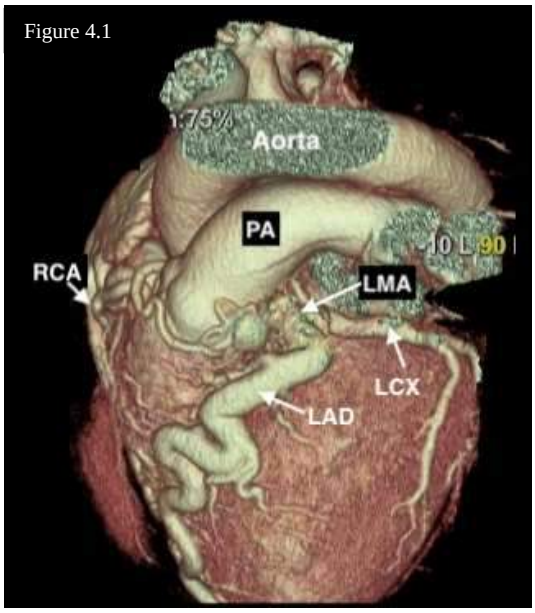
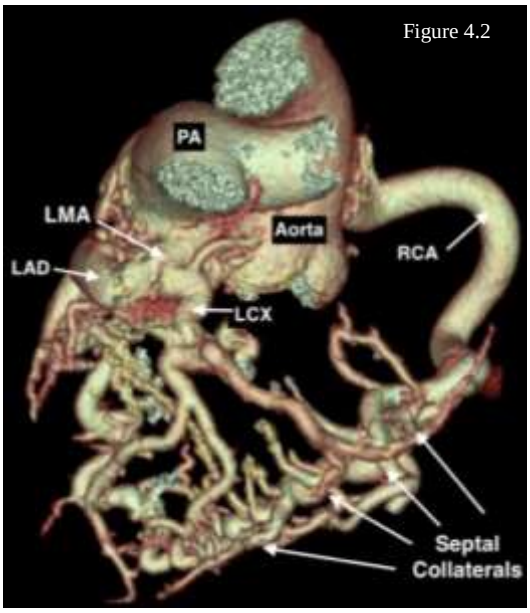
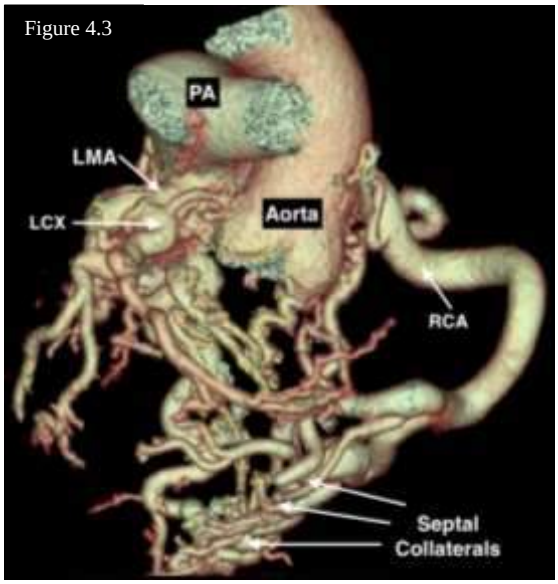



Figure 4.2





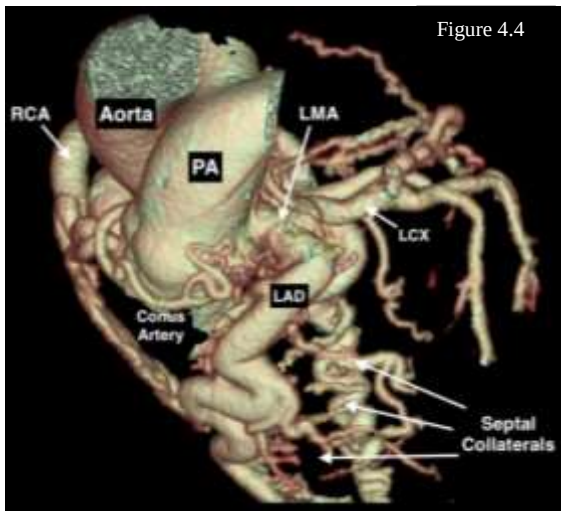




**Figure 4.1, Figure 4.2, Figure 4.3, Figure 4.4: Left Main Artery (LMA) is arising from the Pulmonary Artery (PA), giving rise to Left Anterior Descending Artery (LAD) and Left Circumflex Artery (LCX). Right Coronary Artery (RCA) originating from the Aorta. Multiple septal collaterals arising from RCA and LCA. Conus Artery, the first branch of the RCA (Figure 4.4) is prominent in ALCAPA patients**

As the patient was symptomatic and had dilated LA and LV, which is most likely to be due to chronic ischemia, she was planned for corrective surgery.

## Discussion

CT-CA and Cardiac MRI are some advanced technologies which utilize 3D reconstruction of the coronary artery tree, enabling the distinct visualization of the anatomy of the coronary arteries and their course.

However, CT-CA has its own drawbacks due to exposure to radiation and inability to assess the blood flow despite of producing excellent images. Findings may include direct visualization of the origin of the LCA from the PA, tortuous and dilated RCA, and dilated inter-coronary collateral arteries along the external surface of the heart or within the IVS. Presence and extent of collaterals, quantifying the left to right shunt and measuring end-diastolic pressures may be demonstrated by Angiography and cardiac catheterization.[26],[27] In almost all cases of ALCAPA, even without symptoms, surgical correction is recommended to overcome ischemia, arrhythmias or sudden cardiac deaths which may occur with the progressive increase of left to right shunt. Re-implantation of the LCA to the Left aortic sinus is the treatment of choice. (Takeuchi procedure)[20],[28]

Another option is to place a saphenous vein or an internal mammary graft after completely ligating the anomalous LCA. Benefits of surgical correction include myocardial reperfusion preventing long-term myocardial ischemia and fibrosis, improvement of LV function and probable reduction of the MR.[23],[29] Excellent results may follow from early detection and prompt surgical intervention aimed at restoring a two-coronary-artery circulation system.

## Conclusion

It is evident that patients with anomalous coronary artery origins have varying degrees of presentations, ranging from asymptomatic to inducible ischemia.
CT-CA and 2D echocardiography being important major imaging modalities which identify these anomalies at different stages whereas Cardiac MRI helps to assess the degree of ischemia. In addition to these, cardiac stress tests such as ETT and Dobutamine Stress Echo (DSE) aid in determining the physiological significance of these abnormalities. With the assistance of these imaging and stress testing modalities, decisions on management protocols can be made appropriately.

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# EMPHYSEMATOUS PYELONEPHRITIS- EXPERIENCE AT DGH NUWARA ELIYA

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## Abstract

**Introduction:** Emphysematous pyelonephritis (EPN) is a severe, life threatening infection with gas production in the renal parenchyma, collecting system and perinephric tissue. Rapid progression and deterioration of patients demand early attention and timely management of this condition.

**Methods:** Retrospective study of nine patients presented to us at District General Hospital, Nuwara Eliya for a period of twelve months from October 2021 to September 2022.

**Results:** We demonstrate 9 patients (mean age of 53.4 years; females of 77%) presented to us. Risk factors for EPN were diabetes mellitus (in 100%) and renal stones (in 22%). Clinical presentations include fever (100%), loin pain (88%), vomiting and dysuria. *Escherichia coli* was the most common (88%) urinary isolate. All underwent Computed Tomography (CT) and started on broad spectrum antibiotics. Three (33%) had class 1 EPN and two had class 2 EPN. Class 3A was found in 2 patients and class 3B and class 4 (bilateral EPN) was found in one patient each. Eight patients (88%) had evidence of obstruction (hydroureteronephrosis) in the USS. Besides medical management, eight patients underwent ureteric stenting and three underwent open procedures (two open drainages and one nephrectomy). No mortality occurred in the index cohort.

**Conclusion:** EPN was predominantly noted in female patients and all had a history of diabetes mellitus. Many responded well with minimally invasive procedures apart from medical management and few needed open procedures with no death encountered.

**Key words:** Emphysematous pyelonephritis, DM, *E. coli*, Drainage

## Introduction

EPN is a necrotizing suppurative infection of the renal parenchyma, collecting system and perinephric tissue characterized by gas formation within these structures. [1] It is a life-threatening condition with a high mortality rate and almost always challenges the primary caring team. Diabetes mellitus (DM), obstructive uropathy, vesicoureteric reflux and immunosuppression are the common risk factors for this condition. [2,3] The factors that predispose to EPN in diabetics may include uncontrolled diabetes mellitus, high levels of glycosylated hemoglobin and impaired host immune mechanism. Patients may come with fever, loin pain, vomiting, altered consciousness and shock.[3]

Its clinical significance due to its diagnostic difficulty on clinical grounds, rapid progression and the demand for the timely invasive management strategies in short term necessitates the need for a more vigilant approach in vulnerable population. [4] Outcomes for patients with EPN are variable and depend on status at presentation, comorbidities, radiological class and quality of care.[3] Increasing knowledge on microbiology, pathology, widespread availability of imaging modalities, and multidisciplinary approach of management have picked up EPN at an earlier stage leading to better patient outcome. We present our case series of 9 patients with EPN treated at District General Hospital, Nuwara Eliya from 2021 October to 2022 September.

## Methods

We retrospectively analyzed the medical records of 9 consecutive cases with EPN admitted at District General Hospital, Nuwara Eliya between October 2021 and September 2022. The diagnosis of EPN was based on either initial ultrasound evidence of gas in renal parenchyma, which was confirmed by the subsequent CT scan of abdomen. Patients' demographic, clinical, laboratory and imaging profiles were gathered from their clinical records. Statistical Package for Social Scientists (SPSS version 20) was used to analyze the data, comparing different demographic, clinical and laboratory parameters.

## Results

A total of 9 patients (7 females) were recruited to the study (mean age 53.4 ± 6.7 years; range 42–70) including 7 females (77%). All of them were diabetic and renal stones were found in two patients. The mean delay of presentation to hospital was 3.3± 0.9 days. The common clinical features were fever (100%), loin pain and/or renal-angle tenderness (8 patients; 88%), dysuria (9 patients; 100%), increased urinary frequency (4 patients; 44%) and vomiting (6 patients; 66%). one had mild crepitus in the index loin area and another patient presented with altered consciousness and sepsis. Four patients had been treated for febrile, culture positive urinary tract infection within 2 months prior to the index admission. Neutrophil leucocytosis was noted in all the patients on admission and five patients had thrombocytopaenia. All had high C-reactive protein (CRP) levels. Overall, glycaemic status was poor and all had random blood sugar on admission of >200 mg/dl. Other features were pyuria (9 patients; 100%), albuminuria (8 patients; 88%), hyponatraemia (5 patients; 55%) and microscopic haematuria (7 patients; 77%). Seven cases (77%) were complicated with acute kidney injury. The laboratory profiles on admission are shown in Table 1. The diagnosis of EPN was confirmed by abdominal CT scan they were classified according to Huang and Tseng classification.

*Table -1Initial laboratory profiles of patients on admission*

| Parameter | Mean Value with SD<br>Number of patients |
|---|---|
| Haemoglobin | 9.97 ± 1.67; 7.9-12.3 g/dl |
| Neutrophil Leukocytosis (>11*$10^9$/L) | 9 (100%) |
| Total WCC | 19.47 ± 5.56 ; 7.9-33.2 $10^9$/L |
| Thrombocytopaenia (<150 *$10^9$/L) | 5 (55%) |
| Serum Creatinine | 1.9 ± 1.3 ; 1.3-3.2 mg/dl |
| Blood Urea | 21.6 ± 7.9; 17.6- 49.3 mmol/L |
| Serum Sodium level (<135 mmol/L) | 5 (55%) |
| C-Reactive Protein | 96.9± 40.7; 37-231 mg/L |
| RBS ( >200 mg/dl) | 9 (100%) |

Urine culture isolated the growth of *Escherichia coli* 8 patients (88%), out of which two grew Klebsiella species along with *E.coli*. One patient had sterile urine and blood, but culture of pus collected during stenting revealed growth of *E. coli*. Every culture was reviewed by microbiologist and inputs were taken regarding antibiotics accordingly.

All patients were treated with broad spectrum intravenous antibiotics empirically on admission and other supportive measures including fluid resuscitation and analgesia. All were referred to endocrinologist for blood sugar control and multidisciplinary approach including urologist, radiologist, physician, microbiologist, and endocrinologist was practiced. Initial USS of the abdomen was done in every patient and after review by the radiologist all underwent CT to confirm the diagnosis and assess the extent of the disease. Four patients underwent haemodialysis and maximum haemodialysis sessions needed were four.

According to Huang and Tseng classification, Three (33%) had class 1 EPN and two had class 2 EPN. Class 3A was found in 2 patients and class 3B and class 4 (Bilateral EPN) was found in one patient each. (Figure-1-4) Eight patients (88%) had signs of obstruction (hydroureteronephrosis) in the USS and stented endoscopically. Macroscopically pus drained from the ureteric orifices during stenting. (Figure-5) Patient with Bi lateral EPN had no upper tract dilatation but had intra renal abscess. Percutaneous drainage was done in 6 patients draining pus. (Figure-6)

*Table-2 Comparison of Interventions*

| Surgical interventions | Number of patients |
|---|---|
| Ureteric stenting only | 8 |
| Percutaneous(PC) drainage only | 1 |
| Percutaneous drainage after stenting | 5/8 |
| Open Drainage (Inadequate drainage with stent or PC drainage) | 2 |
| Nephrectomy | 1 |

Two patients needed open drainage of perinephric abscesses and one ended up with nephrectomy. Biopsy of nephrectomy tissue revealed evidence of acute-on-chronic nephritis with micro-abscess formation and renal stones. The patients were hospitalized for 11–29 days. AKI resolved in 6 patients before discharge and there were no mortality encountered in our cohort.

*Table -3 Comparison of Antibiotics and Clinical Outcome*

| | |
|---|---|
| **Antibiotics** | |
| Ceftriaxone | 2 |
| Meropenam | 5 |
| Piperaciliin and tazobactum | 2 |
| Need of Haemodialysis | 4 |
| Return to normal Inflammatory markers on discharge | All |
| **AKI** | |
| Resolved on discharge | 6 |
| Improved on discharge | 3 |
| **Outcome** | |
| Survival | All |
| Death | None |

All the patients were followed up in the clinic with a period of 3 months minimum and all had good recovery.

**Discussion**

Emphysematous pyelonephritis (EPN) is an infective condition of the kidney and perirenal area with high morbidity and mortality. This is characterized by the production of gas within the renal parenchyma, collecting system or the peri-nephric tissues. [(1)] Initially EPN was described as renal emphysema and pneumonephritis. Later, Schultz and Klorfein recommended the term 'Emphysematous Pyelonephritis' (EPN) in 1962. [(4)] Its occurrence is mostly associated with the presence of diabetes mellitus (More than 80% of patients with EPN are diabetes) with a female preponderance (6times commoner than men) as well. [(5)] EPN is caused by glucose fermenting bacteria. In our cohort all had a history of DM and most (7/9) were females. Elderly population is more vulnerable due to their co existing other risk factors and co morbidities. [(5)] The mean age in our cohort was 53.4 years.

Patients with EPN usually present with fever, chills, nausea, and pain in the flanks and abdomen which was reflected in our case series as well. Bilateral involvement of the kidneys is reported to be in only 10% of the cases. [(1)] One patient had bi lateral disease in our cohort. The most common pathological agent is *E. coli*, which is reported to be associated in 69–90% of cases. [(5)] Eight patients in our cohort isolated this organism. The diagnosis is confirmed by radiography. The plain radiograph of the abdomen can show mottled gas in the renal and peri-renal space, but this may occur in only one third of patients. [(6)] The gold standard is the computed tomography of the abdomen, as it will show the presence and the localization of the gas. Huang and Tseng proposed the following classification. [(2)]

*class 1*: Gas presence in the collecting system only (emphysematous pyelitis)
*class 2*: Gas presence in the renal parenchyma without extension to the extra-renal area
*class 3A*: Gas and or abscess presence in the perinephric space
*class 3B*: Gas or abscess presence in the pararenal area
*class 4*: Bilateral EPN of any class or EPN in a solitary kidney

The gravity of the prognosis increases with each class, with class 4 being the worse. [(6)] Patient underwent nephrectomy in our series had class 3B disease.

The therapeutic measures for the management of EPN consist of fluid resuscitation, antibiotics, tight control of sugar, surgical or percutaneous drainage, and nephrectomy if the sepsis is not controlled with other measures. [4] Most of our patients were responded well with medical management and minimally invasive drainage procedures. The type and aggressiveness is guided by the degree of severity of presentation, the predisposing factor, the progress of the clinical situation, and the individual experience and preferences. [5] Most experts advocate an aggressive medical and interventionist approach. Severe cases may require emergent drastic measures such as surgical drainage or even nephrectomy. [5,6] All our patients had good recovery with normalized inflammatory markers on discharge and their recovery period was uneventful. We did not have any mortality.

**Conclusion**

EPN was associated with poor patient survival in the past and regarded as deadly entity. But, with the updated knowledge about the disease and the increased availability of imaging modalities like CT has helped in early recognition of the condition with the anatomical extent. Multidisciplinary approach with broad spectrum antibiotics and other supportive care ensured the halt of progression of the disease. Adequate drainage procedures at an earlier stage lead to a better patient outcome with minimal morbidity and mortality.

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Figures: Figures 1-4 CT images demonstrating EPN. Figure 5- Ureteric stenting with drainage of pus, Figure 6 – PC drainage of pus (Arrows A indicate air in the kidney or peri-nephric tissue and Arrow B denotes stent in Figure 1 PCN tube in Figure 3)

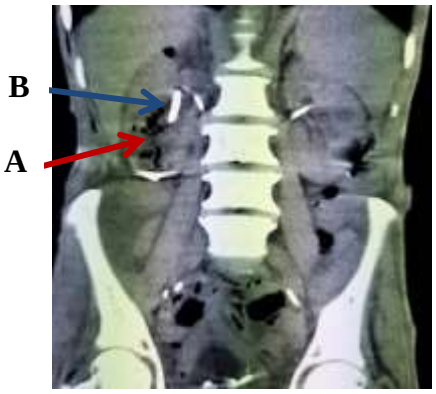


*Figure -1 Coronal View*

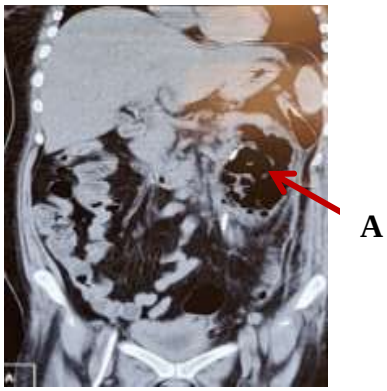


*Figure -2 Coronal View*

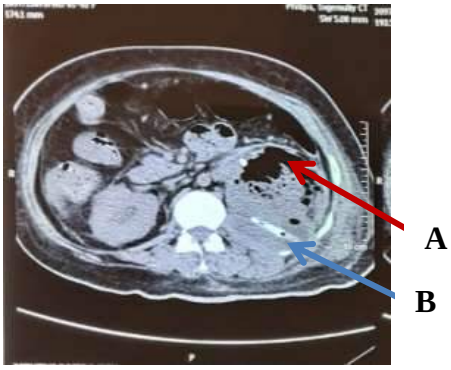


*Figure -3 Axial View*

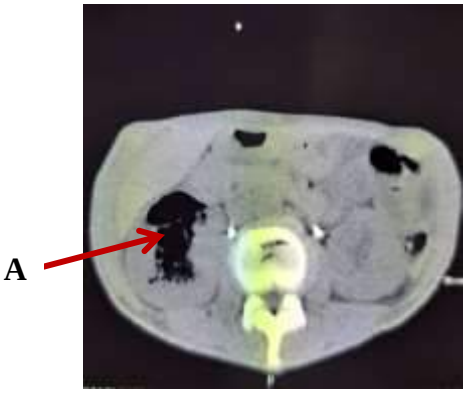


*Figure – 4 Axial View*

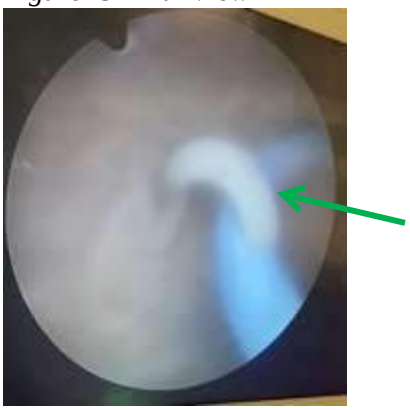

*Figure -5 Draining of pus during ureteric stent*

*Figure-6 PC Drainage of pus*

# CORONARY ARTERY ECTASIA PRESENTING WITH ACUTE INFERIOR ST ELEVATION MYOCARDIAL INFARCTION

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## Introduction

Coronary Artery Ectasia (CAE) is an abnormal finding in coronary angiography and defined as localized, segmental or diffuse dilatation of coronary artery segment to a diameter more than 1.5 times that of adjacent coronary artery. It is well known cause for slow flow and in-situ thrombus formation and patient may presents with complete or partial occlusion of coronary arteries.

In our case report, a 52-year-old male presents with acute inferior ST Elevation Myocardial Infarction (STEMI) and found to have diffuse CAE involving all three main coronary arteries with total distal right coronary artery occlusion during coronary angiography.

## Case History

A 52-year-old male presented with angina at rest for two hours duration. He did not have any history of cardiovascular risk factors for atherosclerosis. On admission, his vital signs were stable and physical examination was unremarkable.

ECG on admission showed ST elevations in inferior leads and ST depressions in LI, aVL and VI to V4 (Figure 01) suggesting acute infero-posterior STEMI.

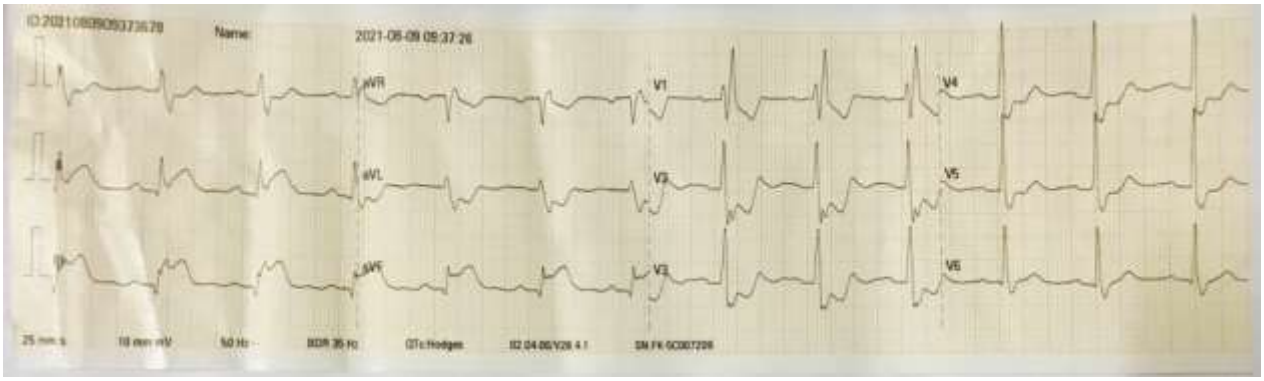


Figure-01 On admission ECG ST elevations in inferior leads and ST depressions in LI, aVL and VI to V4

Coronary angiogram showed diffuse ectasia in LCX, LAD and their branches and RCA showing diffuse large ectasia with distal total occlusion (Figure 02-04).

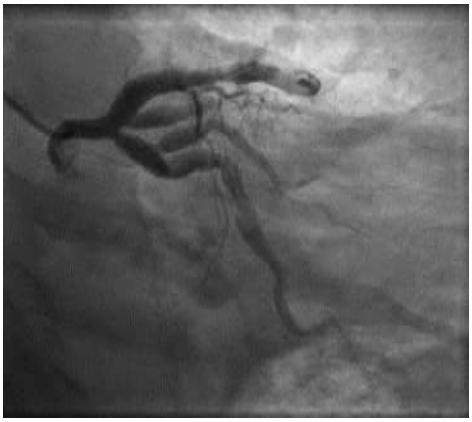

Figure 02: Coronary angiogram showing diffuse ectasia in LCX and its branches

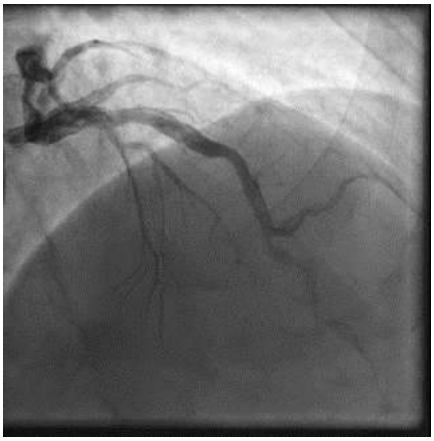

Figure 03: Coronary angiogram showing diffuse ectasia in LAD and its branches

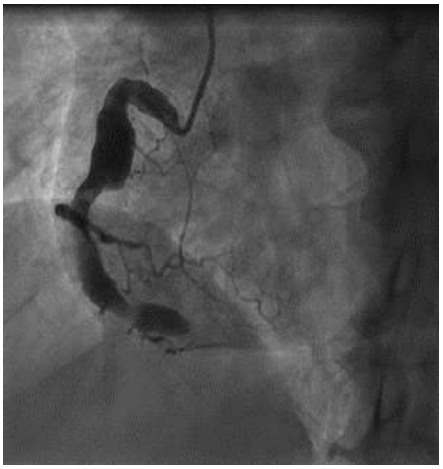

Figure 04: RCA showing diffuse large ectasia with distal total occlusion.

There was no significant flow reversion of RCA despite multiple attempts of balloon dilatation and intracoronary abciximab and coronary angiogram after procedure in Figure 05.

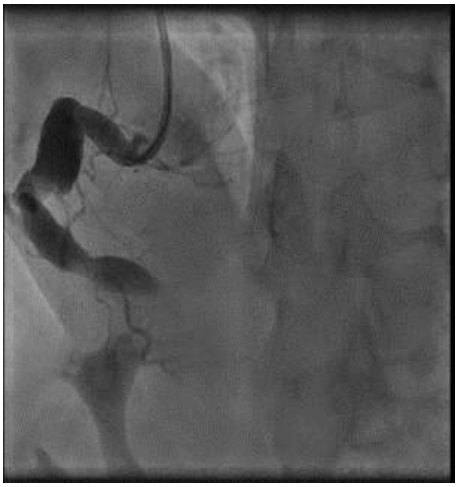

Figure 05: Coronary angiogram after the procedure

**Discussion**

Coronary ectasia is associated with many coronary artery disease risk factors and etiologies while atherosclerosis is the most common.

Coronary angiography in our patient revealed diffuse multi vessel ectasia while larger ectasia measuring 9.9mm in proximal RCA with total distal vessel occlusion. There is no specific management for ectatic coronaries with acute occlusion and same principles for acute coronary syndrome management apply. Long term anticoagulation is recommended as high risk of in-situ thrombus formation in these ectatic vessels [1,2,3].

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# SPONTANEOUS FOCAL CORONARY ARTERY SPASM MIMICKING ACUTE ANTERIOR ST ELEVATION MYOCARDIAL INFARCTION

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## Introduction

Coronary artery spasm is a rare and underestimated course for acute chest pain. It often becomes a diagnostic challenge and it could be easily mistaken for diffuse coronary artery disease. Intracoronary nitroglycerin administration often resolves spasm. We present a case of acute coronary syndrome found to have severe focal coronary artery spasm which resolves completely with intracoronary nitroglycerine.

## Case History

A 33-year-old young female patient presented with angina persisting for six hours. She had similar episode of angina two days back lasting for about half an hour. She didn't have previous history of cardiovascular risk factors.

ECG on admission showed Q with mild ST elevation in V1 and V2 leads ( Figure 01)and ECG on day 2 showed Q with ST elevation and T wave inversion from V1 to V3 and T inversion in V4 (Figure 02).

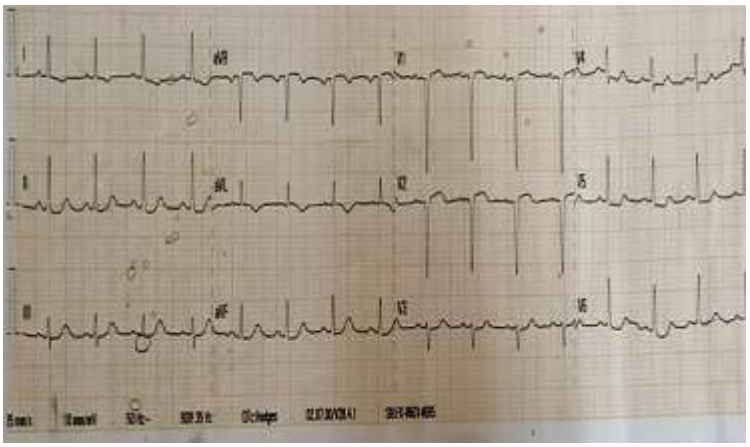

Figure 01: Day 01 ECG (On Admission) showing Q mild ST elevation in V1 – V2

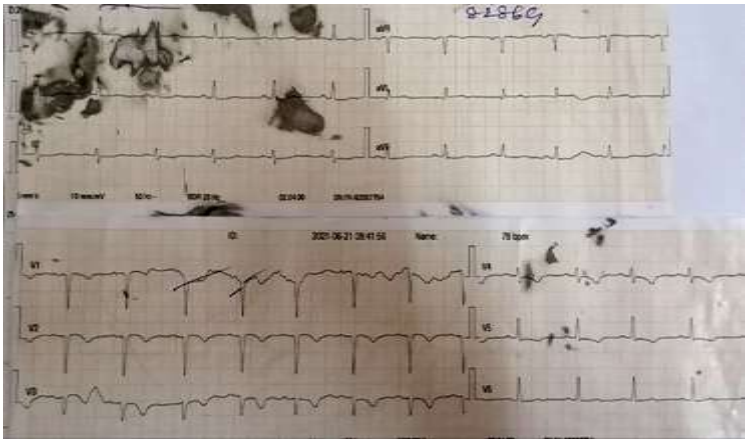

Figure 02: Day 02 ECG showing Q, ST elevation and T inversions in V1 – V3 and T inversion

In the laboratory results, high-sensitive-troponin-I value was 922 ng/L. Other biochemical tests were unremarkable. 2 D echocardiography revealed ejection fraction

of 48% due to basal to mid anterior and anterolateral and anterior apical LV segment hypokinesia. Coronary angiography revealed single, focal severe stenosis in mid LAD (Figure 03, 04 and 05). Other segments of LAD, LCX and RCA were completely normal.

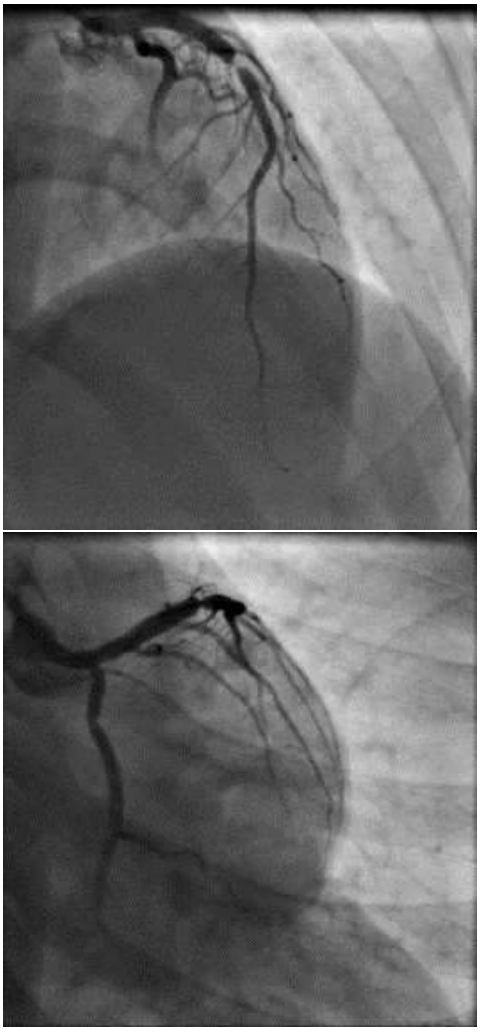

Figure 03 & 04 - Coronary angiogram showing focal proximal vasospasm in LAD with normal LCX

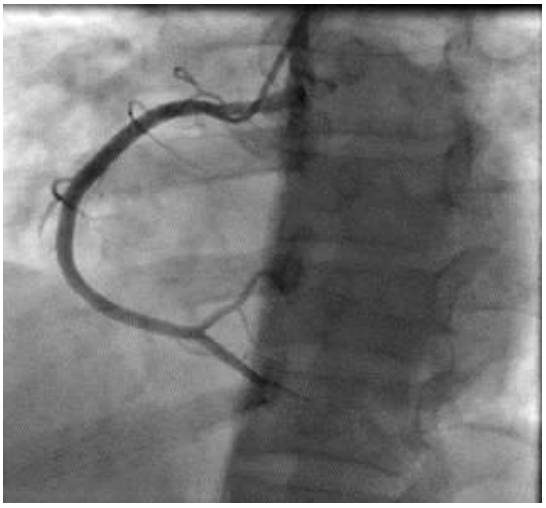

Figure 05 - Coronary angiogram showing normal RCAIntra-coronary nitrate was administered on the basis of the suspicion of vasospasm. Post nitroglycerin injection angiography revealed resolution of the lesion. (Figure 06 and 07).

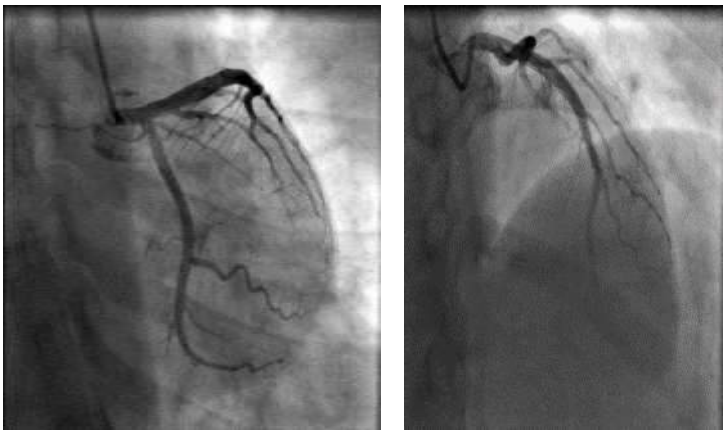

Figure 06 & 07 - Coronary angiogram after Nitroglycerin showing resolution of vasospasm

Nitrates and Ca-channel blockers were added to the routine treatment of the patient, and follow-up coronary angiogram was scheduled in 6 weeks (Figure 08).

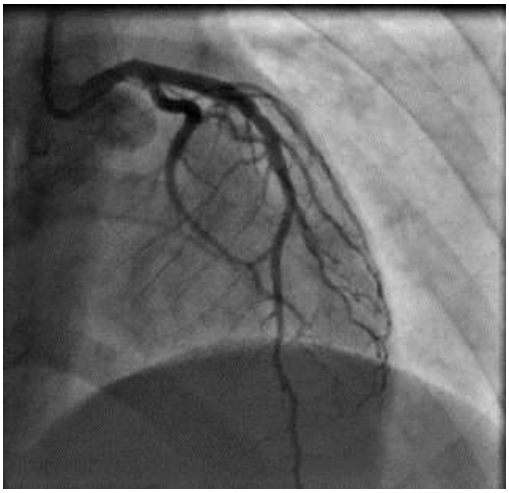

Figure 08 - Follow up Coronary angiogram after 6 weeks showing normal LAD

**Discussion**

Coronary artery spasm is an uncommon presentation of acute chest pain and often causes diagnostic as well as management dilemma. The degree of vasoconstriction ranges from clinically undetectable to complete occlusion and can cause prolonged ischemia. Correct recognition of spasm even during angiogram may not be easy especially in multi vessel spasm [1,2,3]. Coronary vasospasm was suspected in our patient on the basis of the young female without atherosclerosis risk factors and presence of only focal single severe stenosis in LAD. Calcium channel blockers and nitrates are main stay of treatment and beta blockers are generally contraindicated. In our case, intracoronary nitroglycerin administration was able to prevent stent implantation for this patient which has changed the both acute and long term management.

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# A RARE CAUSE OF HEART FAILURE - CARDIAC AMYLOIDOSIS

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**Abstract**
A 61 years old patient presented with symptoms of rapidly worsening heart failure, diarrhea, frothy urine, and easy bruising. His 2D echocardiogram showed the evidence of severe diastolic dysfunction and Global Left Ventricular Strain Pattern showed cherry on the top appearance suggestive of cardiac amyloidosis.

## Case History

61-year-old male who presented with constitutional symptoms together with gradual worsening of exertional shortness of breath (NYHA class III) for one-month duration, with orthopnea & lower leg swelling. He also had watery diarrhea, frothy urine, and easy bruising. On examination, he was found to have macroglossia, ecchymosis (Figure -01), and elevated JVP with pulmonary congestion.

The basic hematological & metabolic panel was normal including the inflammatory markers. The Chest X-ray showed bilateral pleural effusions. He had sub-nephrotic range proteinuria & stool full report showed evidence of inflammatory type diarrhea with blood, with negative cultures. The colonoscopy revealed inflamed colonic mucosa.

His 12-lead electrocardiogram was normal. But his 2D echocardiogram showed non dilated LV with left ventricular ejection fraction of 30 %, poor right ventricular function, left ventricular hypertrophy (IVS 16.5mm, posterior wall 24mm), mild pericardial effusion, severe diastolic dysfunction with bi-atrial dilatation. Global left ventricular strain pattern showed an average of -5.7% with apical sparing (cherry on the top appearance) Figure -02. Although bone-marrow biopsy showed 15-20% of plasma cells. His serum protein electrophoresis & abdominal fat pad aspiration was negative. While the patient was awaiting an endomyocardial biopsy, he passed away due to advanced cardiac failure. Histology from the post-mortem cardiac tissues (Figure -03) were positive for amyloidosis (Figure -04).

## Discussion

Amyloidosis is a rare multisystemic disease that results from the deposition of abnormally folded proteins in organs leading to organ dysfunction. It can present with cardiac or extracardiac manifestations.

Cardiac amyloid deposition causes restrictive infiltrative cardiomyopathy, which carries a high mortality. Light chain (AL) and transthyretin (ATTR) types are the commonest types. ATTR amyloidosis is commonly associated with neurological involvement together with cardiac conduction anomalies. AL amyloidosis is commonly associated with proteinuria, gastrointestinal involvement, purpura together with heart failure. The most likely cause of this patient's cardiac amyloidosis seems to be AL amyloidosis.

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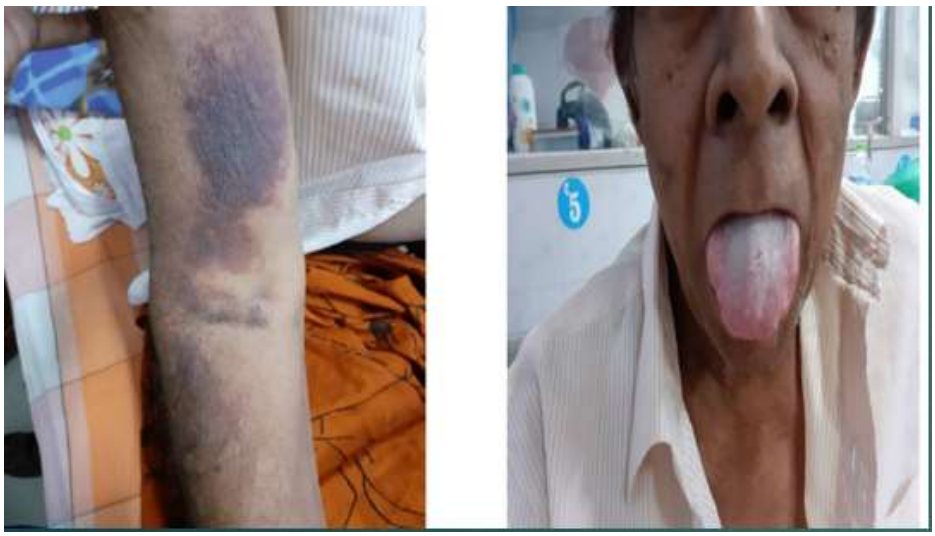
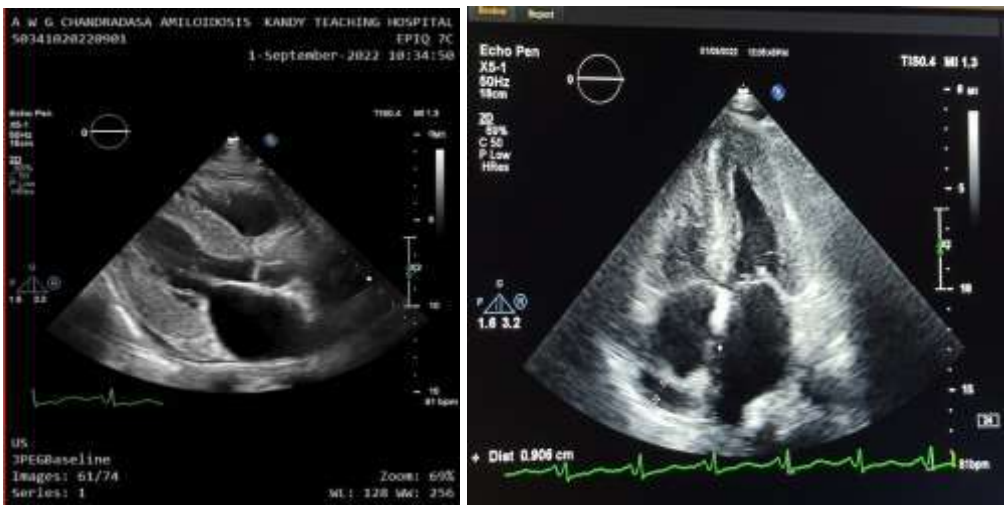


**Figure 01 -** Echymotic patches over venepuncture sites Macroglossia



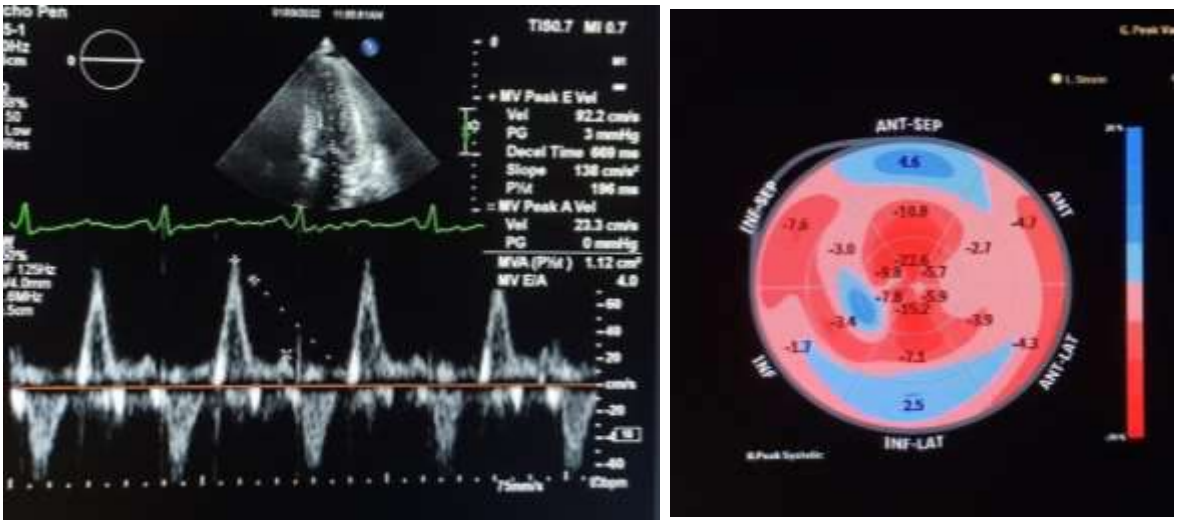



**Figure 02**: 2D Echocardiogram & Global Left Ventricular Strain

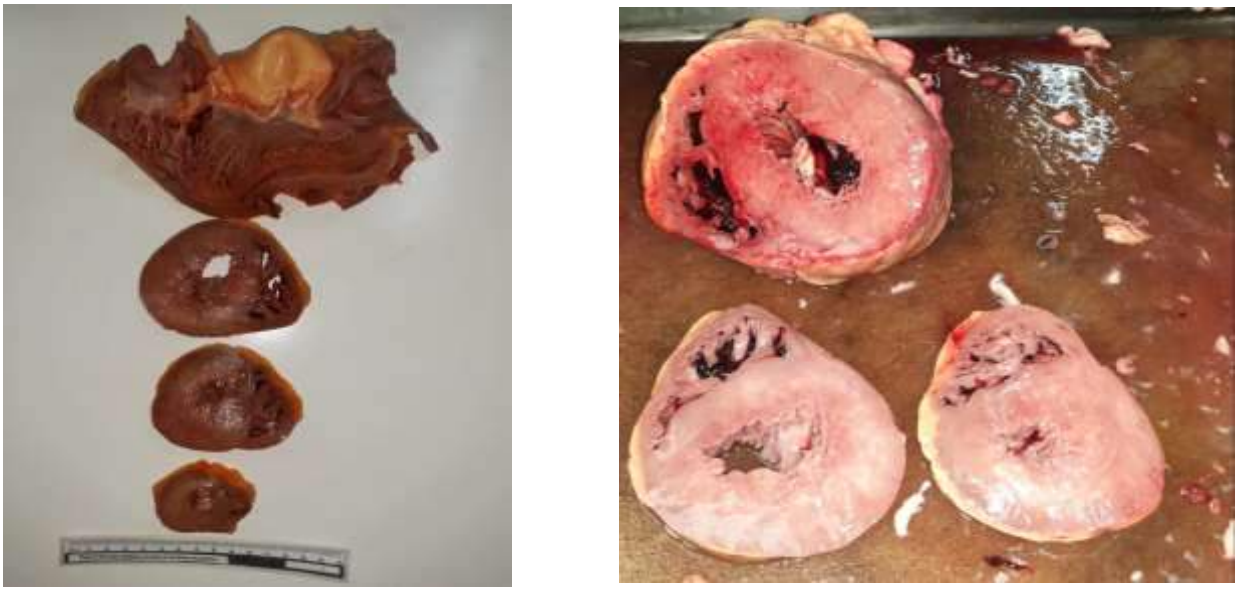


**Figure 03 -** Macroscopic appearance of the post-mortem specimens

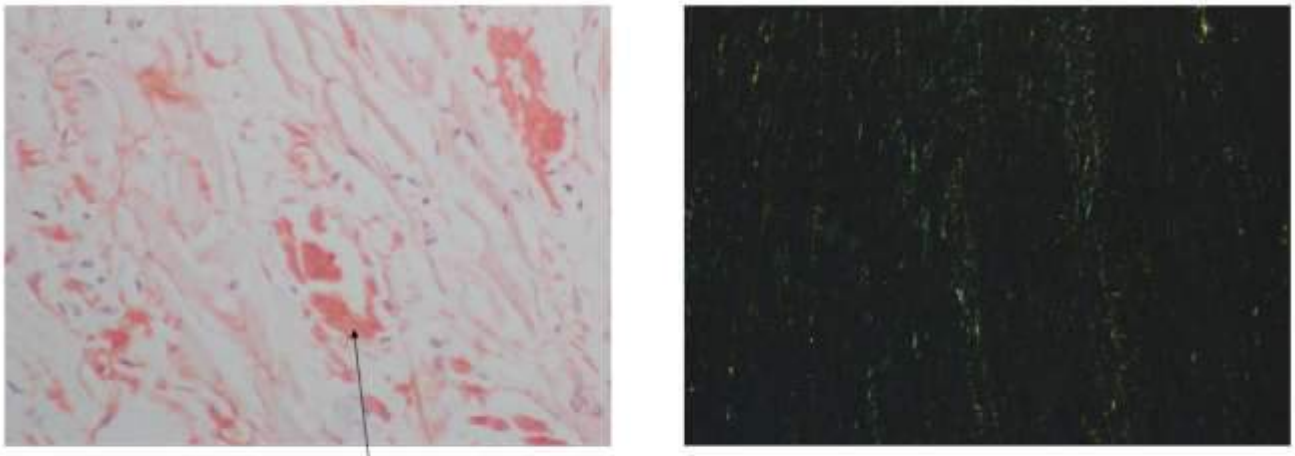



**Figure 04 -** Microscopic appearance of the post-mortem specimens

# METAL RING ENTRAPMENT OF PENIS

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## Case History

A 48-year-old Sri Lankan male bearing a history of a schizoaffective disorder with defaulted follow-up, presented in an afternoon complaining penile strangulation for about 18 hours as a result of failure to remove a metal ring applied at the root of penis. The patient admitted regular self-application of the metal ring with a 3mm thickness to enhance prolonged erection during his bisexual activities. Influence of alcohol and cannabis prevented him from removing the ring after the intercourse previous night. In the morning hours of the day of admission, the patient made multiple failed attempts to remove the ring manually. He visited the hospital in late afternoon when his attempt to divide the ring using a metal grinder failed, partially injuring the engorged penis. The patients' vital parameters were stable. He had a tender, engorged and an oedematous penis which was bleeding due secondary injuries resulted from attempts of self-removal (Figure 1A).

The patient was immediately rushed to the operating theatre and attempts were made to divide the metal ring under general anaesthesia using numerous theatre instruments. Ultimately, a metal grinder from the hospital workshop had to be utilized to divide the metal ring (Figure 1B and 1C). There were no features of reperfusion injury and the patient made an uncomplicated recovery, He received intravenous antibiotics for 48 hours and was discharged after a mental health review. He had normal urinary flow and erectile function at two weeks from the incident.

## Discussion

Penile entrapment or strangulation has been first reported way back in 1755, although this is the first reported case in Sri Lanka according to authors' knowledge. [1] Use of constricting objects like rings and bands made of metal, glass, plastics or latex at the root of the penis for the purpose of self-stimulation, prolonged erection and/or sexual gratification is not currently considered as an illness. However, many times the patients presenting with penile strangulation have been found to be associated with psychiatric disorders. [2, 3] These objects when kept beyond the desired time period would lead to initial venous congestion and subsequent ischaemia of distal penis. Bhat et al. in 1991 has graded these injuries into a scale of five grades, where Grade 1 shows oedema of distal penis without skin ulceration or urethral injury to Grade 5 where there is gangrene/necrosis or complete amputation of distal penis. [2] Goal of management in penile strangulation is for early and safe removal of the constricting foreign body and management of associated injuries. Definitive long term management objectives include follow up to exclude urethral injury/erectile dysfunction, psychological evaluation, education and counseling to prevent future incidents. [3, 4]

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## Figure and Legend

*Figure 1**A** - Presentation with the strangulated penis with secondary injuries 1**B** - Operative removal of the metal ring using a grinder 1**C** - Decongestion of penis immediately following removal of the metal ring.*

# DIFFICULT URETHRAL CATHERIZATION MADE EASY

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The failure to place a Foley catheter is a challenging condition that disturbs the patient and the physician and often needs to be solved immediately. [1] It is one of the most common urological emergencies in daily practice. The main barrier pathologies are urethral strictures and false passages in either sexes, prolapse in females and benign prostatic hyperplasia, prostate cancer, bladder neck contracture and phimosis in males. [2] In some instances, due to the anxiety of the patients and tight external sphincters, a catheter may not be placed normally despite of the normal urethral anatomy. [1]

Difficult urethral catherization (DUC) can end up in significant morbidity to the patient. Cystoscopic assistance might be required in certain failed catheterizations. [2,3] We demonstrate a method to overcome DUC with the use of hydrophilic guidewires without the need for cystoscopes. Apart from the routine instruments for normal catheterization we needed 0.035" hydrophilic guidewire and a large bore needle. (Figure-1A)

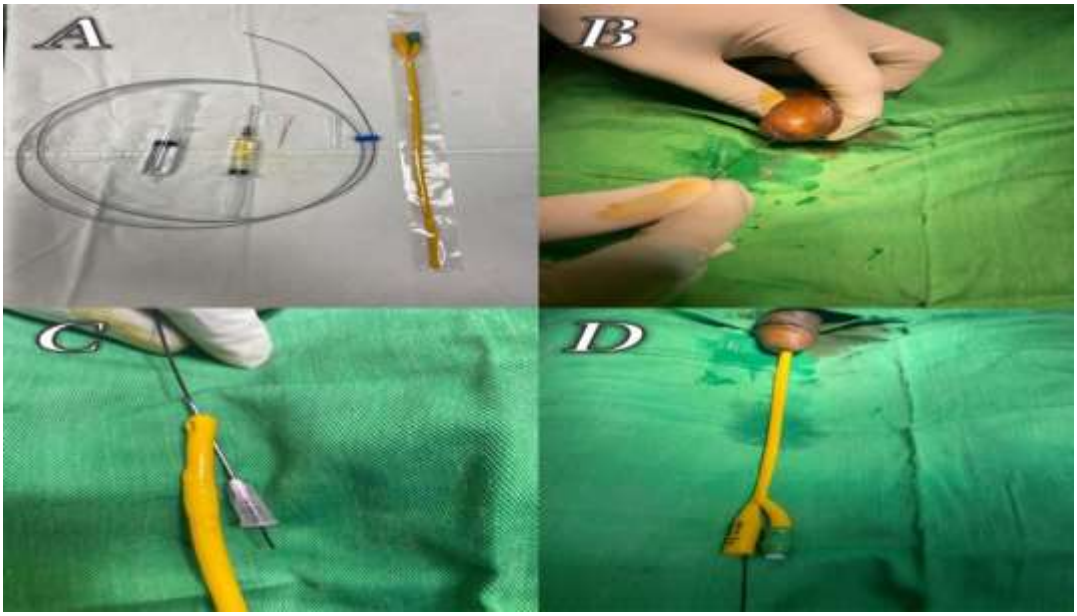
After consenting the patient, genital area was cleaned with betadine. A sterile towel with a hole was placed to expose the penis only. A lubricating gel containing local anesthetic agent applied to the urethra with the help of a syringe and waited for 5 minutes. The saline wetted, soft end of a sterile hydrophilic guidewire was gently advanced through the urethra. (Figure-1B) The open passage in the urethra was searched for by moving the guidewire back and forth at the point it was stuck. When necessary, a gel was applied from the urethral meatus, and the procedure was repeated. If there is any coiling of the guidewire, it was removed and re-attempted. Entry of guidewire into the bladder was confirmed by gentle passage of adequate length of guidewire into the urethra without any coiling. Then the integrity of the catheter bulb was checked in the trolley with inflation and deflation of the balloon. A preliminary puncture was made with the large bore needle from the side hole of the catheter and needle will pierce the center of the tip. Then the stiff end of the guidewire was fed via the needle and taken back into the catheter by withdrawing the needle. (Figure-1C) Then the catheter will be rail-roaded along the guidewire till urine flows. (Figure-1D) After urine flow was observed, the procedure was terminated by inflating the Foley catheter in the bladder and guidewire was removed. The technique described above is an easy, useful, and safe approach in patients with DUC and avoids additional interventions under different conditions.

In DUC, an initial attempt at passing a soft 0.035" guidewire directly into the urethra is quite safe and highly beneficial, and when successful as it gives clear, protected access

to the bladder. A guide wire can be successfully passed in 80% of patients who fail passage of both a coude and a 12 French silicone catheter. [3] At the bedside, a hydrophilic guidewire with a soft tip may be placed into the urethra. The soft tip can be passed beyond obstructions and into the bladder. Any obstruction will either be bypassed by the guide wire, or the wire will reverse direction and pass back out through the urethral meatus.[2,4] If the guide wire is able to pass into the bladder, a catheter can be passed over the wire. Guidewire technique is a less invasive method to reduce the need for additional surgery in difficult urethral catheterizations and studied its outcomes with good success rates under different conditions. [4]

Figure 1:

A - Articles needed, B - Insertion of soft end of guidewire into the urethra, C - Feeding of guidewire into the catheter after a preliminary puncture with needle, D - Rail - roading of catheter

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# A PATIENT WITH ACUTE INFERIOR ST ELEVATION MYOCARDIAL INFARCTION PRESENTS WITH SUBTLE ECG CHANGES AND CONCOMITANT OCCLUSION OF RIGHT AND LEFT CIRCUMFLEX CORONARY ARTERY.

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## Introduction

Twelve lead ECG plays a vital role in diagnosis and predicts the culprit vessel in acute myocardial infarction. ST elevation in inferior leads suggests an acute occlusion of right coronary artery in majority of cases and less commonly in the left circumflex artery. Acute Inferior ST elevation myocardial infarction is diagnosed by the presence of more than 0.1 mV ST elevation in two contiguous leads (LI, LII and LIII) with supportive lateral reciprocal changes. The coronary angiogram on our case revealed concomitant acute occlusion of RCA and LCX which is a rare finding in day-to-day practice.

## Case History

A 71-year-old gentleman with a history of hypertension and COPD presented with angina at rest. On admission, his vital signs were stable and physical examination was unremarkable. ECG on admission and in half an hour is in Figure 1.

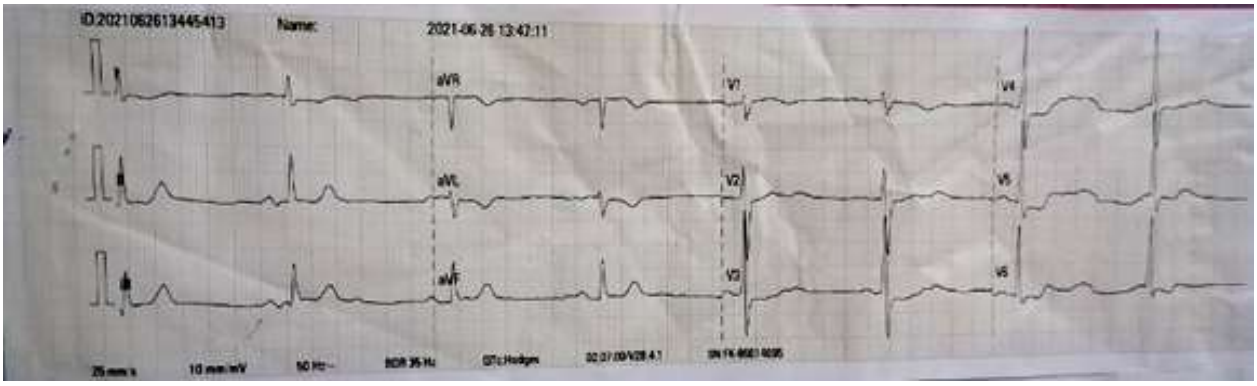


Figure 1a

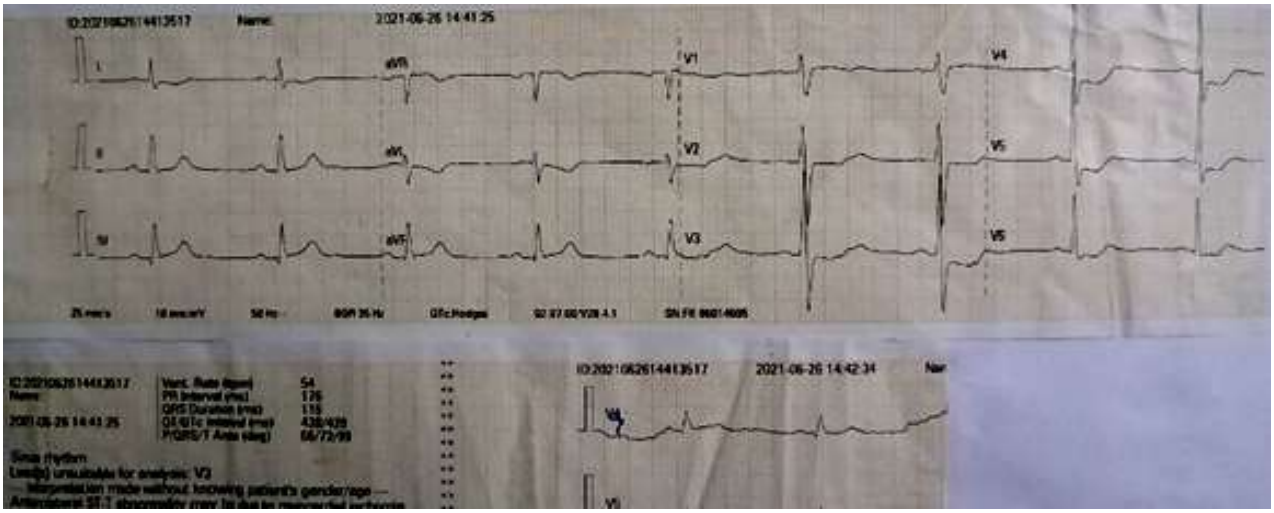

Figure 1b

2D Echocardiogram showed inferior wall hypokinesia. Coronary angiogram showed total occlusion of proximal LCX and RCA, severe stenosis of proximal LAD (Figure 2-4).

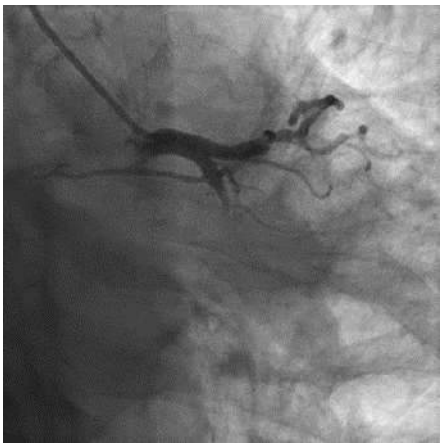

Figure 2- Coronary angiogram showing proximal LCX occlusion

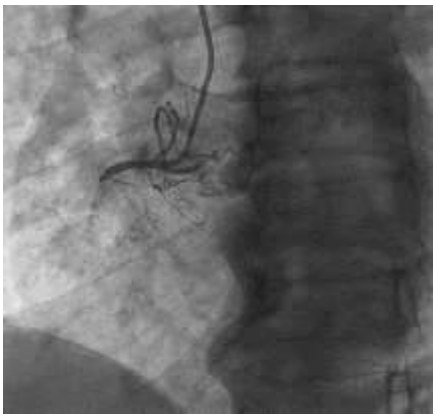

Figure 3 - Coronary angiogram showing proximal RCA occlusion

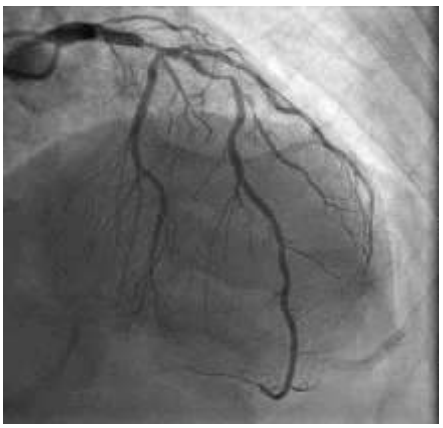

Figure 4 - Coronary angiogram showing proximal LAD severe stenosis

TIMI III flow was established in both vessels following PCI and dominant LCX

lesion was stented (Figure 5 and 6) along with dual antiplatelet therapy.

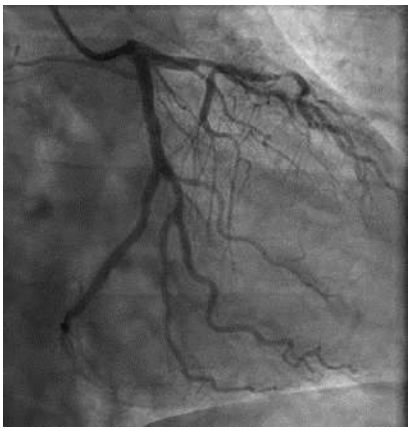

Figure 5 - Coronary angiogram after stenting of proximal LCX lesion

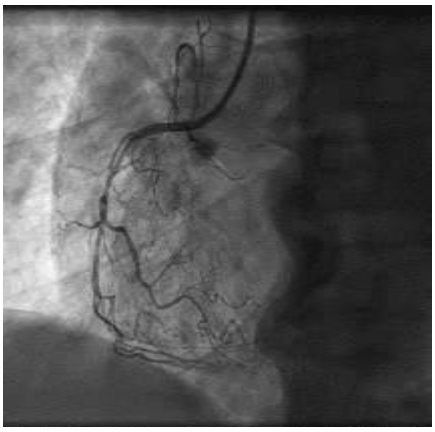

Figure 6 - Coronary angiogram after RCA balloon dilatation

## Discussion

This is a rare presentation of a patient with angina with subtle ECG changes which leads initial diagnostic and management difficulties. The Presence of concomitant occlusion of two culprit arteries (RCA and LCX) is challenging and highlights the importance of restoring the flow in both vessels.

aVL lead is considered as the true reciprocal lead to inferior wall of the heart and the presence of reciprocal changes in aVL lead should be taken specifically when subtle inferior ECG changes are present with strong clinical history.

ST elevation in LIII > LII, reciprocal changes in LI, aVL and right ventricular infarction suggested by V4R lead suggests possible RCA territory where as equal ST segment elevation LII and LIII, ST elevations in lateral leads and absence of reciprocal changes in LI < aVL may suggest possible LCX territory [1,2,3]. ECG changes were not helpful in above case other than presence of reciprocal changes. 2D echocardiogram to see inferior wall motion abnormality and cardiac biomarkers may help to diagnose acute coronary syndrome in early stages of management until coronary angiogram is being performed.

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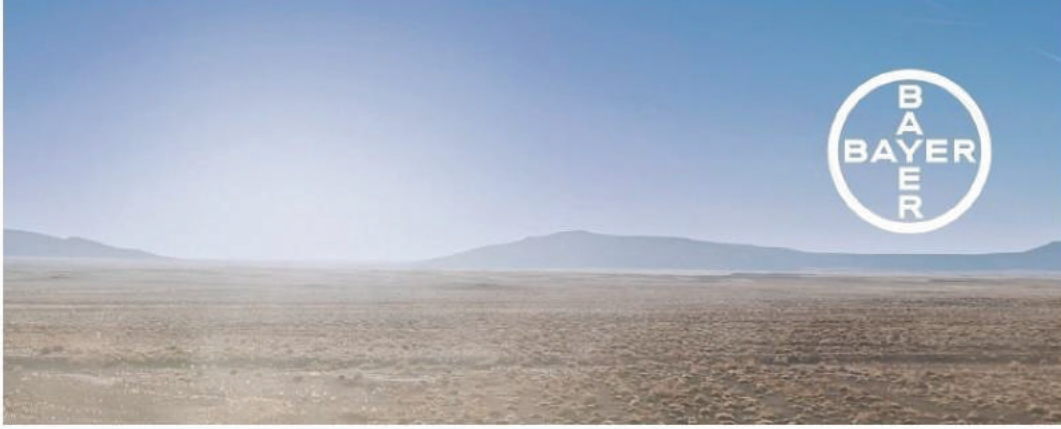


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These events occur rarely. The risk of VTE is highest during the first year of use. The risk of venous or arterial thrombotic/thromboembolic events or of a cerebrovascular accident increases with age; obesity (body mass index over 30kg/m2); a positive family history, if a hereditary predisposition is known or suspected, the woman should be referred to a specialist for advice before deciding about any COC use; prolonged immobilization, major surgery, any surgery to the legs, or major trauma. In these situations it is advisable to discontinue COC use (in the case of elective surgery at least four weeks in advance) and not to resume until two weeks after complete remobilization; smoking; dyslipoproteinemia; hypertension; migraine; valvular heart disease; atrial fibrillation. Other medical conditions which have been associated with adverse circulatory events include diabetes mellitus, systemic lupus erythematosus, hemolytic uremic syndrome, chronic inflammatory bowel disease (Crohn's disease or ulcerative colitis) and sickle cell disease. Other conditions require precautions are women with hypertriglyceridemia, sustained clinically significant hypertension, women with hereditary angioedema, acute or chronic disturbances of liver function, Crohn's disease and ulcerative colitis. **Drug Interactions:** Effects of other medicaments on Yaz: microsomal enzyme-inducing drugs e.g. phenytoin, barbiturates, primidone, carbamazepine, rifampicin, and possibly also oxcarbazepine, topiramate, felbamate, griseofulvin and products containing St. John's wort, antibiotics (except rifampicin and griseofulvin), also HIV protease (e.g. ritonavir) and non-nucleoside reverse transcriptase inhibitors (e.g. nevirapine), these drugs can cause contraceptive failure. Women on treatment with any of these drugs should temporarily use a barrier method in addition to the COC or choose another method of contraception. *Effects of COCs on other medicaments* Oral contraceptives may affect the metabolism of certain other drugs. Accordingly, plasma and tissue concentrations may either increase (e.g. cyclosporin) or decrease (e.g. lamotrigine), an increase in serum potassium in women taking Yaz **Pregnancy and Lactation:** Yasmin is not indicated during pregnancy. If pregnancy occurs during treatment with Yasmin, further intake must be stopped. Lactation may be influenced by COCs as they may reduce the quantity and change the composition of breast milk. Therefore, the use of COCs should generally not be recommended until the nursing mother has completely weaned her child. **Undesirable Effects:** Following are the common (> 1/100, <1/10) undesirable effects; emotional lability, depression/ depressive mood, Decrease and loss of libido, migraine, nausea, breast pain, unscheduled uterine bleeding, genital tract bleeding not further specified. **Presentation:** Calendar-pack containing 21 tablets YSM APIv2_09 Mar 2021 (CDS 17_23 Mar 2017)

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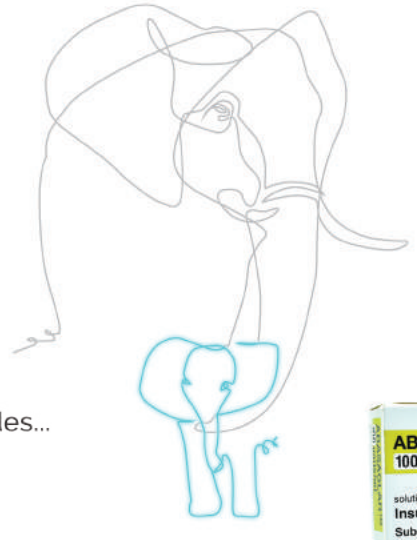

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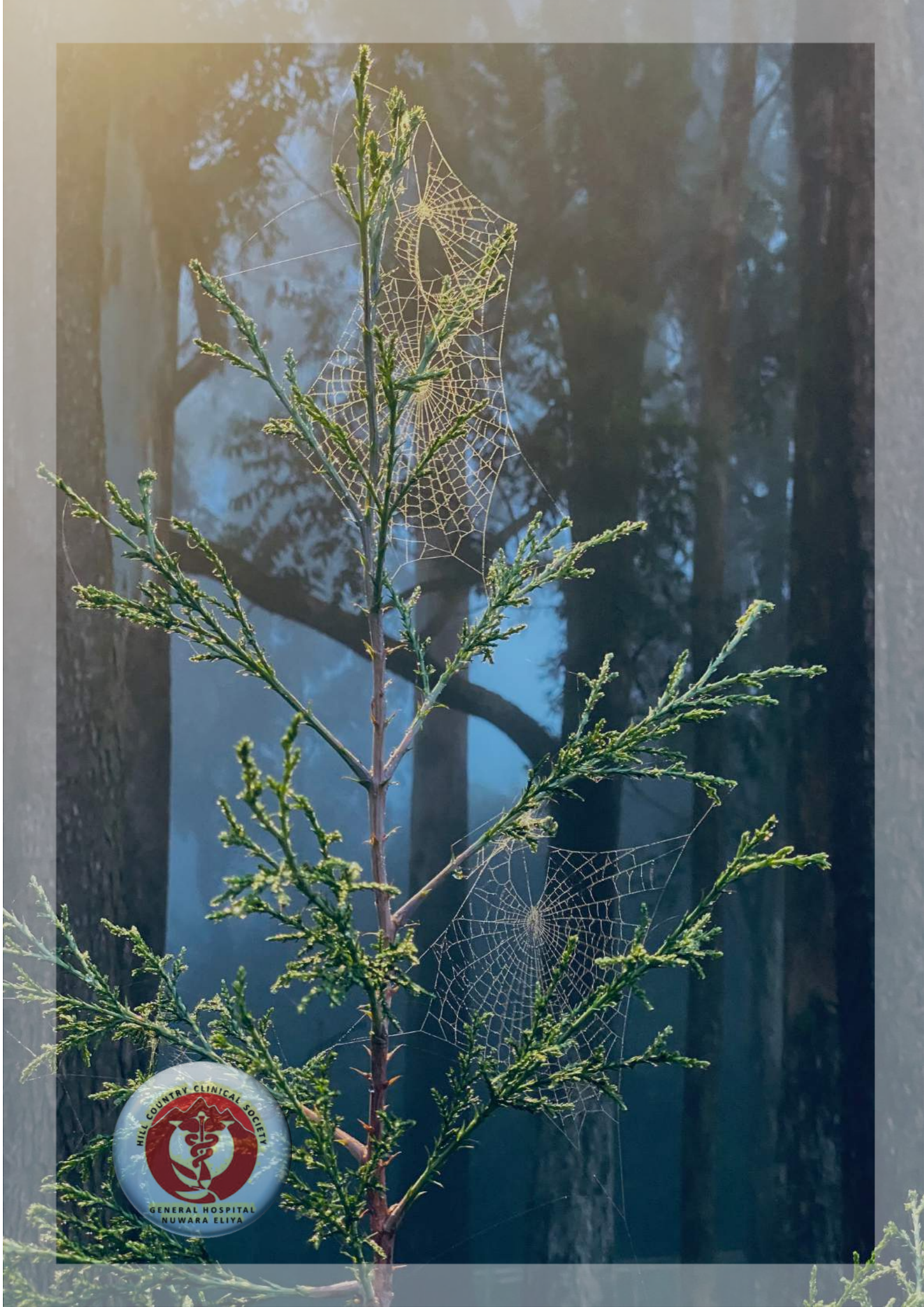
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