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HealthNews DIGEST

AUGUST 2024

Dr. Sherbaz Bichu

CEO & Specialist Anaesthetist
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The 24th edition of "HealthNews Digest" is here!

On behalf of the leadership, I am delighted to congratulate all on the 24th edition of the HealthNews Newsletter. It's been two years since we started this, and I am delighted to share the overwhelming success of these newsletters. Without our doctors' remarkable dedication and steadfast commitment to patient care and medical excellence, this would not have been possible.

Throughout the year, you have diligently worked to present our distinguished readers with various topics, from innovative treatments to perceptive articles. Your commitment, enthusiasm, and expertise have been instrumental in the success of HealthNews.

As we embark on this edition, I express my gratitude to everyone for sharing their profound knowledge. Let's keep innovating, exploring, and collaborating together to produce content that leads the way in the dissemination of medical knowledge.

Dr. Ramanathan V

Medical Director
Aster Hospitals & Clinics, UAE

As the Medical Director for Aster Hospitals and Clinics, it gives me great pleasure to welcome you to the 24th edition of our HealthNews Digest Newsletter. Congratulations on this milestone achievement!

These newsletters are nothing short of extraordinary due to the collective effort and dedication of our exceptional team of doctors. I genuinely appreciate all contributors for making these newsletters a huge success.

Your commitment to compassion, resiliency, and patient care is evidence of the enormous impact that healthcare providers can have on their patients' lives. The efforts you put forth to diagnose and treat complex cases and support patients during their most vulnerable times are incredibly admirable.

Together, let's achieve even more incredible feats to further our aim of improving the medical field.

Dr. Rajesh Vijay Kumar
Internal Medicine (Specialist)
Al Nahda, Sharjah

Dr. Akta Trivedi
Neurology (Specialist)
Al Qusais, Dubai

Dr. Jayakumar B. Kannan
Gastroenterology (Consultant)
Al Muteena, Deira, Dubai

An unusual case of Bleeding - "Hereditary Haemorrhagic Telangiectasia (HHT)" managed successfully at Aster Clinic, Al Nahda, Sharjah

PRESENTATION

- 38-year-old female
- Presented with:
  - Long-standing history of fatigue
  - Frequent intravenous iron transfusions over the last 5 years that helped in improving the symptoms
  - Intermittent dark-coloured stools and epistaxis
  - Skin lesions over both lower limbs
  - Unilateral headache for 3 months that worsened from last 1 week; no side predilection and throbbing, moderate in intensity and increasing in frequency
- Family history of similar symptoms (mother and two siblings). The patient's mother is also taking frequent iron injections to improve the symptoms

FINDINGS

On Examination:

- Telangiectasias over both legs
- No pallor, oedema or lymphadenopathy
- Conscious and oriented with normal systemic examination

Blood Investigations:

- Haemoglobin: Normal
- Mean Corpuscular Haemoglobin Concentration (MCHC): Low
- Serum Iron: Normal
- Serum Ferritin: Normal
- Renal Function Test: Normal
- Liver Function Test: Normal
- Prothrombin Time: Normal
- Activated Partial Thromboplastin Time (aPTT): Normal

A gastroenterology opinion was sought to identify any gastrointestinal cause for her condition.

Ultrasonography Abdomen:

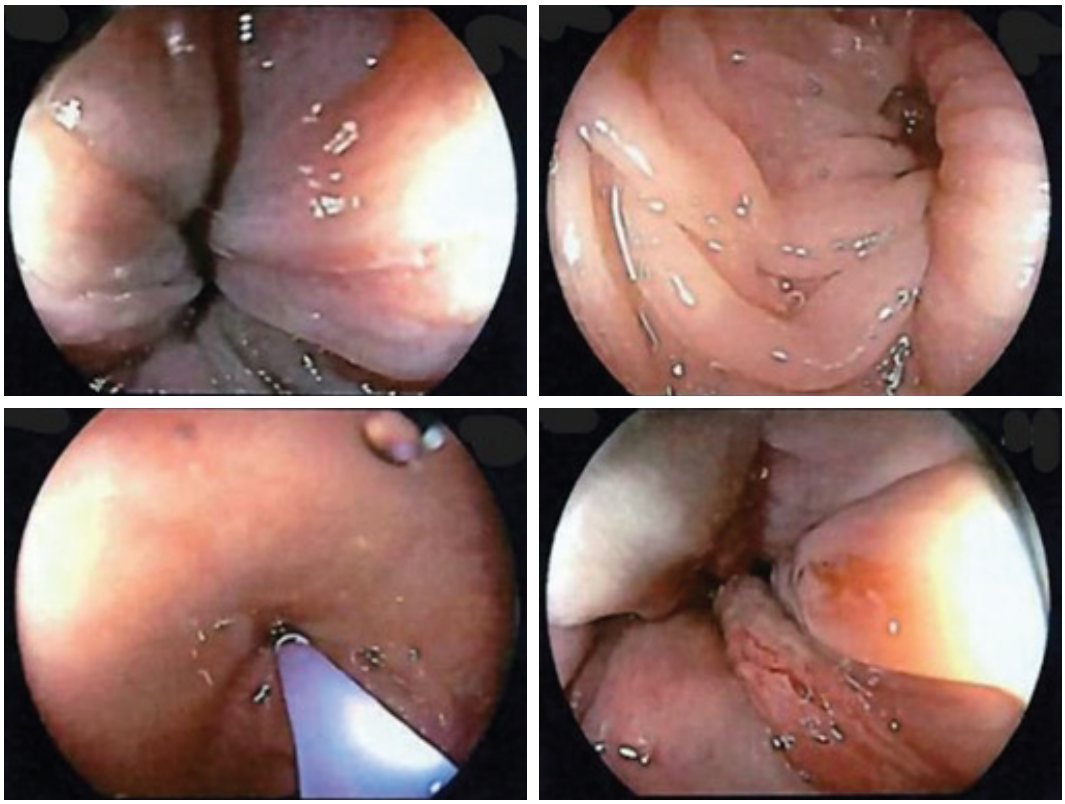
- Grade 1 diffuse fatty infiltration of the Liver

Upper Gastrointestinal Endoscopy:

- Fundal gastritis and duodenitis
- Gastric biopsy showed chronic inactive gastritis and duodenal biopsy was negative for Celiac disease or malignancy

Colonoscopy:

- Patchy colitis, proctitis, posterior skin erosions and haemorrhoids
- Colonic biopsy showed mild chronic inactive colitis



Colonoscopy Images

The patient was later referred to the Neurology Department to evaluate her headache.

On Neurological Examination:

- CNS examination: Normal
- Brain imaging was done to rule out cerebral arteriovenous malformations or any other cerebrovascular lesion.

MRI Brain Plain:

- Normal study
- The patient responded to symptomatic medical management.

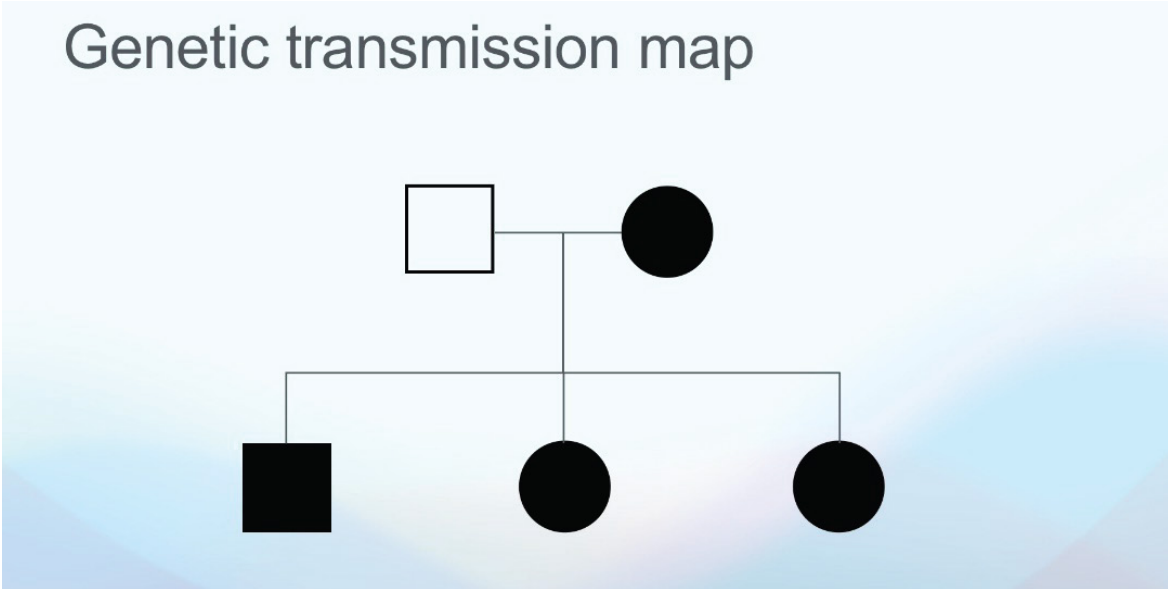
DIFFERENTIAL DIAGNOSIS:

The differential diagnosis based on the initial evaluation included the following bleeding disorders:

- Von Willebrand Disease (Epistaxis)
- Hereditary Haemorrhagic Telangiectasia (Epistaxis with Telangiectasias)
- Congenital platelet function disorders like Bernard-Soulier Syndrome, Glanzmann Thrombasthenia, Hermansky-Pudlak Syndrome and Scott Syndrome
- Acquired Platelet function disorders due to drugs, uremia, liver disease, and dysproteinaemia.

EVALUATION AND TREATMENT

An evaluation of the genetic expression of this disease in her family was done. The inheritance pattern was shown to be a dominant transmission, as depicted in the genetic diagram below:



- Among the differential diagnoses stated above, only HHT (Hereditary Haemorrhagic Telangiectasia) and vWD (von Willebrand Disease) were transmitted in a dominant (autosomal) pattern.
- Moreover, as her clinical features partially satisfied the criteria for HHT, she was advised to proceed with genetic testing for herself and her brother.
- Her brother's genetic testing showed positive heterozygosity for ENG (p.Arg93d*), one of the three genes (ACVRL1, ENG, SMAD4) for HHT.
- Her genetic testing showed positive heterozygosity for ENG (p.Arg93Ter).
- Hence, she was diagnosed with Hereditary Haemorrhagic Telangiectasia (HHT).

She was managed conservatively with symptomatic treatment, including nasal decongestants, multivitamins and lifestyle modification. There were no conditions requiring admission to the hospital or active intervention. However, she was advised to follow up on a regular basis and monitor her bleeding symptoms closely.

DISCUSSION

Hereditary Haemorrhagic Telangiectasia (previously known as Osler-Weber-Rendu disease) is a rare autosomal dominant disorder with multiple mucocutaneous telangiectasias and visceral arteriovenous malformations. It has an estimated frequency of 1-20 cases/100,000 (1) that affects blood vessels throughout the body (causing vascular dysplasia) and results in a tendency for bleeding. The prognosis varies depending on the severity of symptoms; generally, it is good as long as the bleeding is promptly recognized and adequately controlled.

Most patients with HHT experience epistaxis, mucocutaneous telangiectasias, gastrointestinal bleeding, and a tendency to develop iron deficiency anaemia secondary to blood loss (2).

In addition, arteriovenous malformations (AVMs) frequently affect the pulmonary, hepatic, and/or cerebral circulations, demanding knowledge of the risks and benefits of screening and treating patients with these complications.

The diagnosis is made clinically based on the Curaçao Diagnostic Criteria for HHT, established in June 1999 by the Scientific Advisory Board of the HHT Foundation International, Inc (3). The four clinical diagnostic criteria are as follows:

- Epistaxis
- Telangiectasias
- Visceral lesions
- Family history (a first-degree relative with HHT)

The HHT diagnosis is classified as definite if three or four criteria are present, possible or suspected if two criteria are present, and unlikely if fewer than two criteria are present.

Screening family members for signs of HHT is reasonable and should include a complete history, physical examination, chest radiography, and arterial blood gas testing (with measurement of the shunt fraction). Indications for intervention in HHT vary according to the site of involvement and presentation (4).

In mild cases, no treatment is necessary. In more severe cases, treatment consists of managing bleeding via medical and surgical options, as well as surgical management of AVMs and further sequelae.

Individuals with HHT present to a wide range of clinicians spanning medical, surgical, general practice disciplines, and emergency departments, most of whom lack appreciation of the full range of consequences of the diagnosis of HHT for patients and their families (5).

The estimated incidence of CNS involvement in HHT patients is 10-20%. Migraine headaches occur in 13-50% of patients with HHT. Although the reason is unclear, the headaches are more prevalent in patients with pulmonary AVMs. Other neurologic involvement occurs in 8-12% of patients with HHT (6). A history of headache, seizures, and focal neurologic symptoms (e.g., paraplegia or paralysis) may be presenting symptoms.

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Iron Deficiency Anaemia in Children

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INTRODUCTION

Iron deficiency (ID) is a significant global health issue characterized by plasma ferritin levels below 12 µg/L, in the absence of infection or inflammation (1). More specifically, ID is defined by a transferrin saturation (TS) of less than 10% or serum ferritin below 15 µg/L (1). Iron deficiency anaemia (IDA) is often used as a proxy indicator for iron deficiency (ID), which affects 20 to 25% of preschool children worldwide (1). It is widely believed that most cases of ID result from poor dietary iron intake and low iron bioavailability (1). The progression of iron deficiency occurs in three successive stages: depletion of iron stores, iron-deficient erythropoiesis, and ultimately, the development of IDA (1). IDA is diagnosed when anaemia is confirmed to be due to iron deficiency (1). This condition adversely affects children's psychomotor development and cognitive abilities, making it the most prevalent micronutrient deficiency in young children globally (1).

This article discusses the causes, risk factors of IDA, how to diagnose patients, and the treatment options available.

SYMPTOMS OF IDA

Symptoms of IDA can vary widely and become more pronounced as hemoglobin levels drop below 7-8 g/dL (2). A key indicator of IDA is pallor, especially noticeable on the palms, nail beds, and conjunctiva (2). Common symptoms include palpitations, shortness of breath during exertion, headaches, ringing in the ears, dizziness, and fainting (2). In infants, IDA may manifest as breath-holding spells, disrupted sleep, and developmental delays (2). The condition also affects neurodevelopment, impacting cognition (2). Additional signs of IDA include decreased taste sensation, brittle hair and nails, nail changes, and angular cheilitis (2). Rarely, spoon nails, blue sclera, and dysphagia (Plummer-Vinson syndrome) may occur (2).

Older children may experience fatigue, cognitive impairment, and a sensation of cold (2). In severe cases, symptoms can escalate to tachycardia, heart failure, loss of appetite, and signs of pica, which can also be an early symptom (2).

CAUSES OF IDA

Following conditions may cause IDA:

a) Prematurity:

Newborns experience a physiological decrease in hemoglobin levels known as physiologic anaemia, with premature infants being particularly vulnerable (3). Premature newborns can reach a hemoglobin nadir of 7-8 g/dL within 4-6 weeks after birth, while full-term newborns reach a nadir of 9.5-11 g/dL at 9-12 weeks (3). Due to anaemia of

prematurity (AOP), around 90% of extremely low birth weight (ELBW) newborns will require at least one packed red blood cell (PRBC) transfusion (3). The etiology of AOP is multifactorial, including impaired intrauterine iron transport, low erythropoietin (EPO) production, and a shortened RBC lifespan (3). Additionally, maternal conditions such as obesity, gestational diabetes, hypertension, and placental insufficiency contribute to the high prevalence of iron deficiency in preterm infants (3). Frequent blood sampling in critically ill premature infants, inadequate nutrition, infections, and other non-laboratory blood losses also play significant roles in the development of AOP (3).

b) Neuromotor Disorders:

IDA is prevalent in children with swallowing impairments because they often require gavage feeding of liquid or semi-liquid diets, which may exclude essential nutrients (4). Furthermore, conditions such as gastroesophageal reflux are common and can lead to chronic bleeding when complicated by esophagitis (4).

c) Gastrointestinal Diseases:

Conditions like celiac disease, Helicobacter pylori infection, chronic inflammatory bowel disease, pernicious anaemia, chronic blood loss from cow's milk protein intolerance, Meckel's diverticulum, and intestinal parasitosis cause refractory IDA (4). Early testing for celiac disease and H. pylori is crucial in refractory cases (4).

d) Extra-intestinal Blood Loss:

Among additional sources of blood loss outside the gastrointestinal tract, menorrhagia can also aggravate blood loss in adolescents (4). Rarely, such bleeding may also be attributed to Von Willebrand disease, a congenital coagulation disorder characterized by mild to absent bleeding tendencies, which can often go undetected until menarche (4).

e) Refractory IDA:

Refractory IDA is characterized by a lack of response to oral treatment, even after ensuring that factors affecting therapy adherence such as inadequate dosage, the timing of administration, type of iron used, duration of treatment, and the presence of inflammation or infection have been meticulously ruled out (4). These cases typically represent secondary forms of IDA, and identifying the underlying cause can be complex (4).

f) Iron-Refractory Iron Deficiency Anaemia (IRIDA):

In cases of unexplained IDA, consideration should be given to IRIDA, which is genetically linked to mutations in the TMPRSS6 gene (4). Individuals with this mutation exhibit elevated levels of hepcidin, a hormone that inhibits iron absorption in the gastrointestinal tract (4). This condition explains why affected individuals do not respond to oral iron therapy (4). The TMPRSS6 gene likely encodes a protease responsible for reducing the synthesis of hepcidin.(4).

RISKS OF IDA

IDA typically peaks during childhood due to a mismatch between dietary iron intake and the increased demand for iron during rapid growth (4). Other contributing factors include bleeding, and disorders affecting intestinal iron absorption (2). Breast milk provides higher iron absorption (50%) compared to cow's milk (10%), which can lead to IDA if cow's milk is consumed excessively (2). Infants with low birth weight or perinatal blood loss are at higher risk due to inadequate iron stores (2). Delayed cord clamping (1-3 minutes) reduces ID risk, while early clamping (<30 seconds) increases it (2).

Additional risk factors include a history of prematurity, lead exposure, exclusive breastfeeding beyond four months, and transitioning to whole milk and complementary foods without iron-fortified options (4).

DIAGNOSIS OF IDA

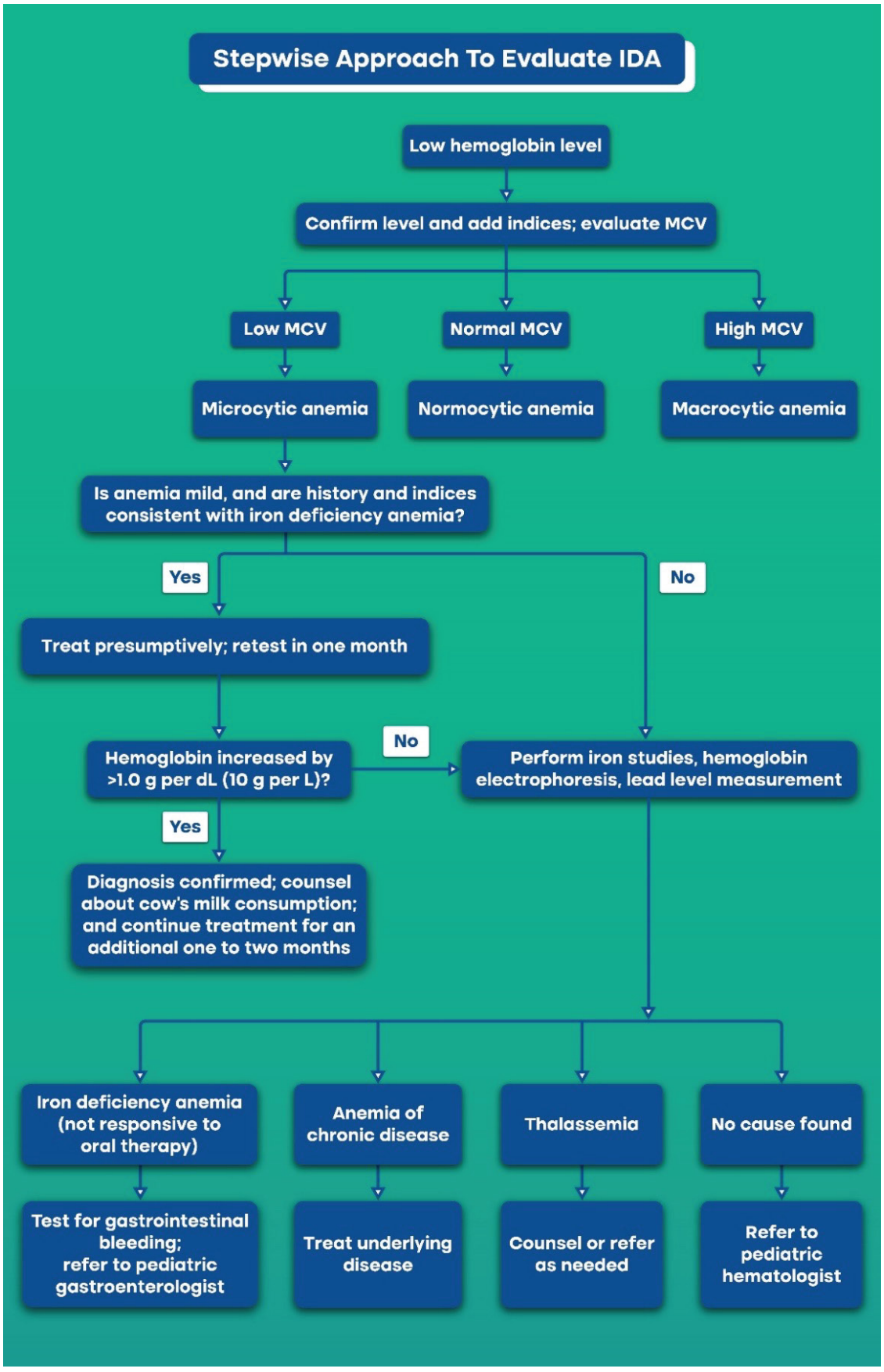

Figure 1: Evaluation of IDA (5)

a) Medical History and Physical Examination:

Physical examination takes notes changes in paleness of the face, palms, conjunctiva, and oral mucosa, as well as signs such as alopecia, brittle and rough nails, koilonychia, and angular stomatitis. Patients may also report a sore tongue, dry mouth, and atrophy of the lingual papillae (6).

b) IDA Screening and Testing:

Screening for IDA is debated due to financial considerations (7). In low-income countries, widespread iron supplementation may be more practical than hemoglobin screening (7). The WHO recommends screening children when the prevalence of anaemia exceeds 5% (7). The American Academy of Paediatrics (AAP) suggests universal screening at one year of age, while the Centers for Disease Control and Prevention (CDC) advises targeted screening for high-risk children, such as premature infants, those in poverty, refugees, and those on low-iron diets (7).

In the diagnosis of iron deficiency anaemia, a combination of clinical evaluation and laboratory tests is essential. The following figure 2 outlines the key diagnostic criteria used to identify iron deficiency anaemia in paediatric patients.

Screening Parameters of IDA

Diagnostic Parameter	Range/Value
Hematologic Characteristics of IDA	
Hemoglobin (infants & toddlers 6 months-5 years)	<11 g/dL
Hemoglobin (children 5-12 years)	<11.5 g/dL
Hemoglobin (adolescent females >12 years)	<12 g/dL
Hemoglobin (adolescent males)	<13 g/dL
Red Cell Distribution Width (RDW-CV)	>15%
Reticulocyte Production Index	<0.5%
Hemoglobin A2	1.5-3.2%
Platelet Count (Thrombocytosis)	>400,000/µL
Hemoglobin Content of Reticulocytes (CHr)	<26 pg
Biochemical Characteristics of IDA	
Serum Iron	<40 µg/dL
Serum Ferritin	<20 µg/L (<100 µg/L if functional iron deficiency or sequestration is present)
Serum Transferrin	>400 µg/dL
Transferrin Saturation	<20%
Serum Zinc Protoporphyrin (ZnPP)	>40 µmol/mol
Soluble Transferrin Receptors (sTfR)	>35 nmol/L

Figure 2: Hematologic and Biochemical Features in the Diagnosis of IDA (7)

COMPLICATIONS OF IDA

IDA in children, especially mild cases, often goes unnoticed by parents and healthcare providers since it may not show obvious symptoms (8). However, even mild anaemia can harm a child's growing body and brain (8). The body adjusts to lower hemoglobin levels, but the effects on growth and cognitive development persist (8).

IDA significantly affects the developing brain of a child, children diagnosed with IDA often perform poorly in memory, reading, language, and math skills (8). Some of these cognitive deficits may persist despite iron treatment. Anaemia disrupts the mental processes crucial for learning and understanding, impacting abilities such as reasoning, memory retention, judgement, and problem-solving (8). These effects develop slowly and may not be immediately noticeable (8).

Furthermore, IDA leads to physical weakness, decreased appetite, reduced ability to engage in activities like playing and running, and decreased overall productivity (8). It weakens the child's immune system, making them more susceptible to infections and potentially increasing the risk of complications such as stroke, thrombosis, congestive heart failure, and even mortality (3,8).

TREATMENT AND MANAGEMENT

a) Dietary Therapy:

Non-pharmacological management focuses on optimizing dietary iron intake (6). Iron is available in two forms: heme iron, which is more bioavailable and derived from animal-based sources, and non-heme iron, which is found in plant-based foods (6). Combining iron-rich foods with vitamin C-rich sources is recommended to enhance intestinal iron absorption (6). However, dietary therapy alone is insufficient for treating iron deficiency anemia because the inherent iron content in foods has limited bioavailability (6). In contrast, supplementation with ferrous iron compounds provides a more readily absorbed source to address IDA (6).

b) Oral Iron Products:

IDA treatment typically begins with oral iron supplementation due to its effective, safe, and cost-effective nature (4). The commonly used preparations include ferrous sulfate, ferrous gluconate, ferrous fumarate, ferrous acetate, ferrous ascorbate, and ferric citrate (4). Other formulations include heme iron polypeptides, carbonyl iron in oral suspensions or tablets combined with folic acid, zinc, vitamin C, or vitamin B12, and iron chelates like ferrous bis-glycine chelate and ferric tris-glycine chelate (7). Additionally, iron hydroxide polymaltose complex, sucrosomial iron, and other liposomal forms are available (6).

Treatment for iron-deficiency anaemia involves administering one 60 mg iron tablet daily for school-age children and two 60 mg iron tablets daily for adolescent boys, alongside counseling on dietary iron intake (9).

c) Parenteral Iron Therapy:

Parenteral iron therapy is recommended for patients who are unable to tolerate or do not respond to oral iron supplements, have difficulty swallowing iron preparations, or experience persistent malabsorption issues (7). Parenteral therapy allows precise calculation of the required iron dose to normalize hemoglobin levels and replenish iron stores, and causes fewer gastrointestinal side effects (7).

Second-generation compounds such as low molecular weight iron dextran, iron saccharate, and ferric gluconate are widely used as they don't cause anaphylaxis, although they require multiple infusions (4). Third-generation compounds, including ferric carboxymaltose and iron isomaltoside, offer the advantage of high-dose administration in a single infusion (4). Only ferric carboxymaltose is approved for paediatric use in patients over age 14 (4).

d) Treatment of Underlying Conditions:

Various health conditions can contribute to anaemia in children, necessitating specific actions to address these underlying causes (10). Preventing and treating malaria, schistosomiasis, and other infections caused by soil-transmitted helminths are essential steps (10). Managing chronic diseases such as obesity and digestive problems is also important (10). Treat heavy menstrual bleeding and menstrual abnormalities to prevent blood loss (10). Delaying umbilical cord clamping after childbirth, ensuring it is not done earlier than one minute, can also prevent IDA in infants (10). Additionally, treating inherited red blood cell disorders like sickle-cell disease and thalassemia is critical in managing anaemia in children (10).

e) Monitoring and Follow-Up:

Once normalized, Hb concentration and red cell indices should be monitored at regular intervals: every 3 months for the first year, again after an additional year, and subsequently if anaemia symptoms reappear (11).

PREVENTION OF IDA THROUGH NUTRITION

Educating parents and caregivers about iron-rich foods and proper dietary practices is crucial for preventing IDA. Foods rich in iron include (4,12).

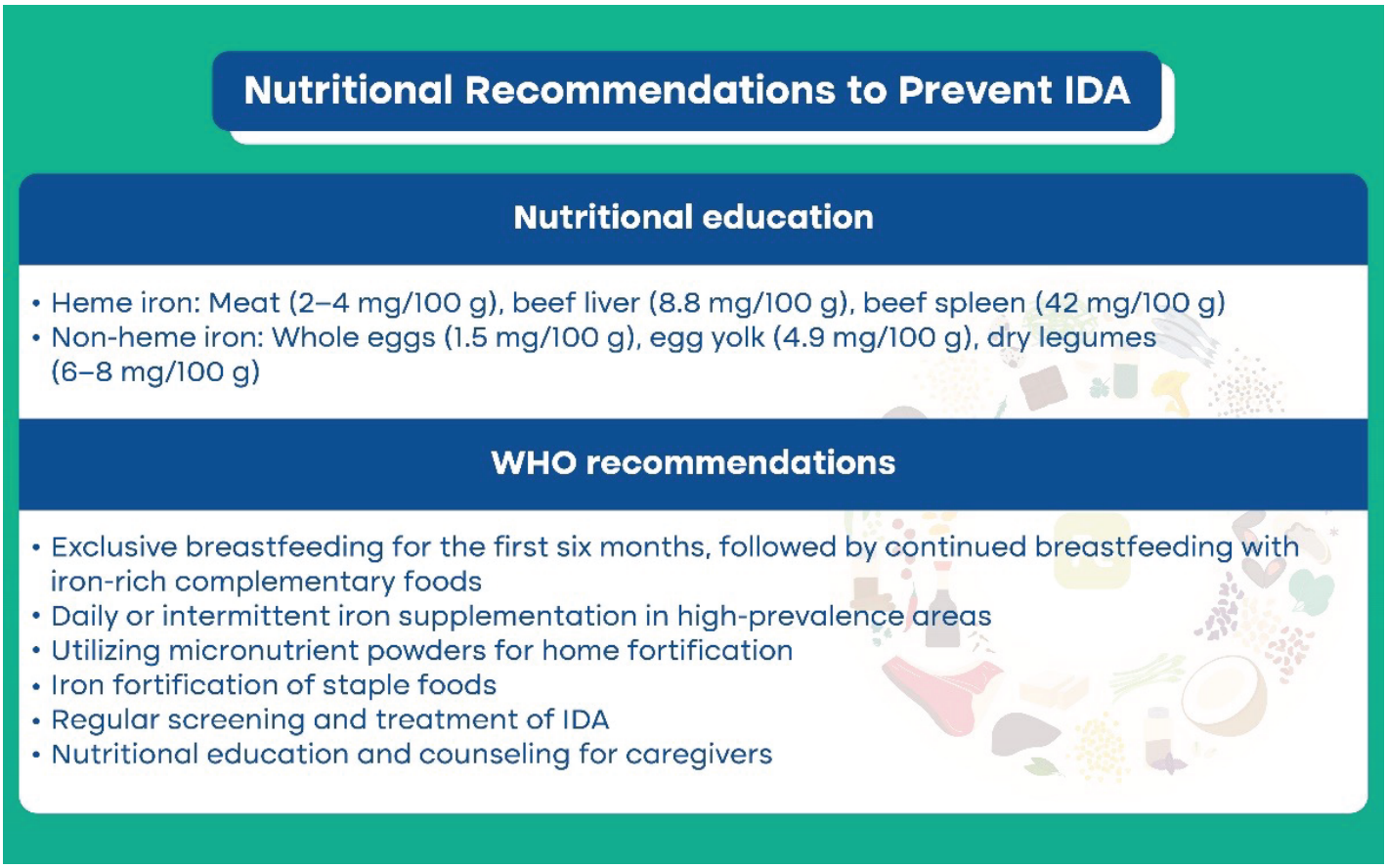

Figure 3: Preventing IDA with Iron-Rich Food (4,12)

Key Highlights

- IDA globally impacts children, affecting cognitive development and growth (1).
- Common symptoms in children involve pale skin, irritability, fatigue, rapid heartbeat, sore or swollen tongue, and cravings for non-food items like ice or dirt – pica (2).
- Treatment strategies include dietary therapy, oral iron supplementation (e.g., ferrous sulfate) or intravenous iron therapy (e.g., ferric carboxymaltose) for patients unable to tolerate oral supplements or requiring rapid correction (4–6,10).
- IDA can be prevented by educating caregivers about iron-rich foods, promoting breastfeeding supplemented with iron-rich foods after six months, and fortifying staple foods with iron to mitigate the risk of IDA (4,11).

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Successful Endoscopic Removal of an Impacted Foreign Body from the Terminal Ileum at Aster Hospital, Mankhool

Dr. Gireesh Gopinathan
Gastroenterology (Specialist)

PRESENTATION

- 25 year old male
- Prior surgical history of Colectomy with anastomosis 18 years back
- No family history of medical illness
- Admitted to ER with:
  - Complaints of severe right-side lower abdominal pain for 1 day with more pain over the right lower quadrant
  - Patient gave a history of ingestion of mutton bone the previous day

FINDINGS

During Examination:

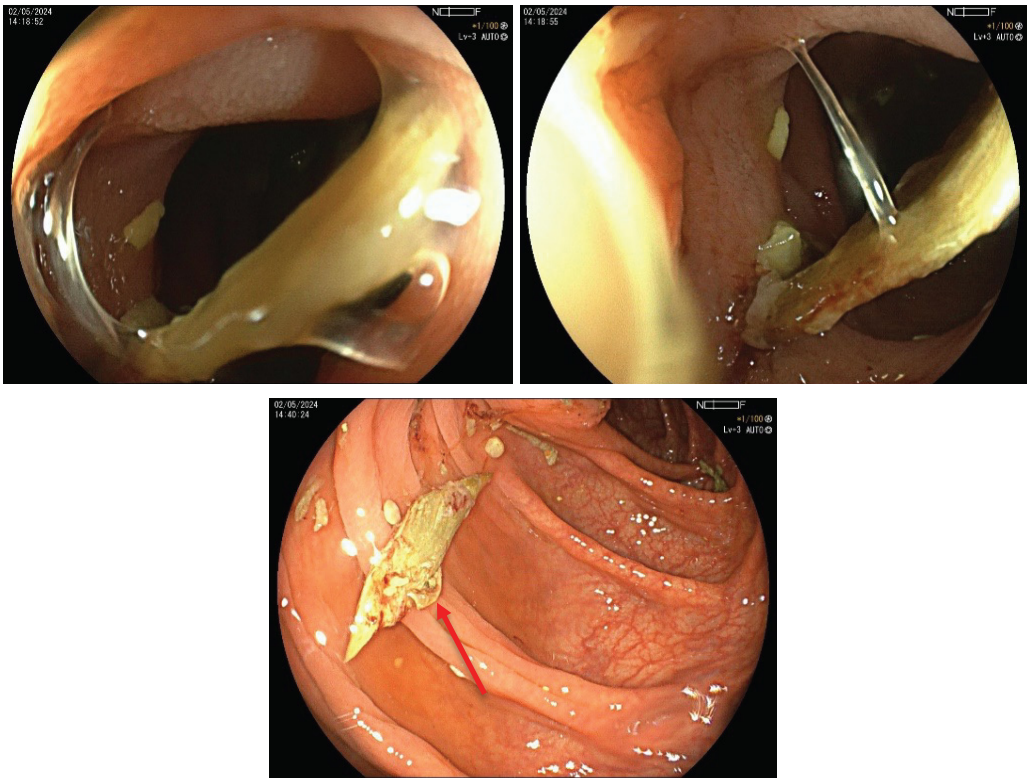
- Stable vitals
- Soft abdomen
- Mild tenderness at the right lower quadrant

The patient was admitted and evaluated. **CECT Abdomen** was done that showed:

- A linear horizontal hyperdense structure of 24 mm in length and 3 mm in width in the terminal ileum suggested a foreign body.
- The foreign body was abutting the terminal ileum's anterior and posterior walls.
- A foci of intramural air was seen in the posterior wall.
- Edematous long segment thickening of the terminal ileal loop associated with mesenteric fat stranding.
- A pocket of loculated air was seen along the medial wall of the ileum approximately 8-10 cm proximal to the Ileocecal (IC) junction, likely to be sealed off perforation.

DURING PROCEDURE

- Colonoscopy was attempted after explaining the risk to bystanders.
- It was seemingly hard to navigate the colon because of the adhesions throughout the length.
- Managed to reach the cecum and IC valve, but intubating the T-ileum and sustaining the position was difficult.
- 1.5 cm sharp bone was seen impacted in the terminal ileum.
- One end was disimpacted and removed with rat tooth forceps.



Colonoscopy images showing the Mutton Bone impacted in the Terminal Ileum

POST PROCEDURE

There was no evidence of perforation, and the patient was started on a gradual oral soft diet the next day. His condition was stable at the time of discharge, and his abdominal pain had improved.

DISCUSSION

Accidental ingestion of a foreign body is often encountered in clinical practice; however, intestinal perforation rarely develops as the swallowed foreign body usually advances through the gastrointestinal tract without any problems and is excreted with faeces. Only 1% of ingested objects result in gastrointestinal system perforation. These materials are generally long and sharp-edged. Perforation most often occurs in the terminal ileum and recto-sigmoid region and is characterised by angulations (1).

We usually encounter such impacted foreign bodies in children, and there have been multiple case reports where they were retrieved endoscopically and surgically. However, it is quite rare in the adult population, yet there are a few case reports (2).

In this case, the endoscopic approach was used to remove a large, sharp-edged, impacted mutton bone with the support of endoscopy staff and technicians.

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Diabetic Kidney Disease (DKD): Comprehensive Management and New Therapies

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INTRODUCTION

Diabetic kidney disease (DKD) is the primary cause of chronic kidney disease and/or end-stage kidney disease, affecting approximately 40% of individuals with diabetes (1). Common clinical manifestations of DKD include albuminuria, weight gain, peripheral edema, nocturia, nausea, anemia, and hypertension (1). The pathophysiology of DKD involves various pathways, such as hemodynamic, metabolic, and inflammatory processes, with risk factors classified as either modifiable or non-modifiable (2).

For an accurate diagnosis of DKD, the assessment of estimated GFR and albuminuria, along with consideration of clinical features such as the presence of diabetic retinopathy (DR) and duration of diabetes, is essential (1). Effective management of DKD is multifaceted and typically involves a combination of primary prevention, lifestyle modifications, and pharmacotherapy (1).

This article will provide detailed information on the diagnosis, pharmacological treatment, and emerging therapies for managing DKD.

DIAGNOSIS OF DIABETIC KIDNEY DISEASE

The clinical evaluation of DKD involves assessing several parameters, including the duration of diabetes mellitus (DM), presence of elevated blood pressure, presence of diabetic retinopathy in addition to measurements of eGFR and albuminuria (3).

DKD is clinically identified by a persistent urinary albumin-to-creatinine ratio ≥ 30 mg/g and/or a consistent decline in eGFR to less than 60 mL/min/1.73 m^2 (3). To confirm albuminuria or a low eGFR, at least two abnormal measurements must be taken three months apart (3).

Other causes of kidney diseases must be excluded first before diagnosing DKD. These can be suggested by severe proteinuria, presence of dysmorphic RBCs in urine, red or white cell casts, or other systemic diseases that can cause kidney disease e.g SLE. In these cases kidney biopsy can be considered.

Recently, it has been noted that diabetic kidney disease can present with a phenotype not associated with proteinuria, which is considered an atypical presentation but increasingly recognized.

According to the American Diabetes Association (ADA), DKD screening should be conducted at least once a year for patients with type 1 diabetes mellitus (T1 DM) starting five years after diagnosis as well as for all patients with type 2 diabetes at their initial visit, as patients with type 2 Diabetes are often asymptomatic and can have DKD at the time of DM diagnosis (4).

MANAGEMENT OF DKD

The two primary objectives in managing DKD are: preserving renal function to mitigate the risk of ESKD and reducing the likelihood of cardiovascular events and mortality (3). Additionally, individuals with DKD are at an elevated risk of developing diabetic retinopathy, neuropathy, and foot ulcers, necessitating close monitoring for these associated complications (3).

Lifestyle Modifications:

According to guidelines from the ADA and Kidney Disease: Improving Global Outcomes (KDIGO), adopting lifestyle modifications such as achieving a healthy body weight, regularly engaging in physical activity (≥150 minutes/week), and abstaining from tobacco use is beneficial (4,8). Additionally, these guidelines recommend dietary interventions that include a low-sodium intake (KDIGO: <2,000 mg/day; ADA: 1,500 to <2,300 mg/day) to help manage blood pressure and reduce cardiovascular risks (4,8). All these are considered to be a foundation for lifestyle modification.

Based on clinical characteristics, first-line drug therapies are added to lifestyle changes targeting glycemic control (metformin, SGLT2i), blood pressure control (RAS blockade) and dyslipidemia (Statins). If targets are not achieved using these first-line drugs, additional drugs can be added to achieve those targets including GLP1 receptor agonists and nonsteroidal mineralocorticoid receptor antagonists (ns-MRA).

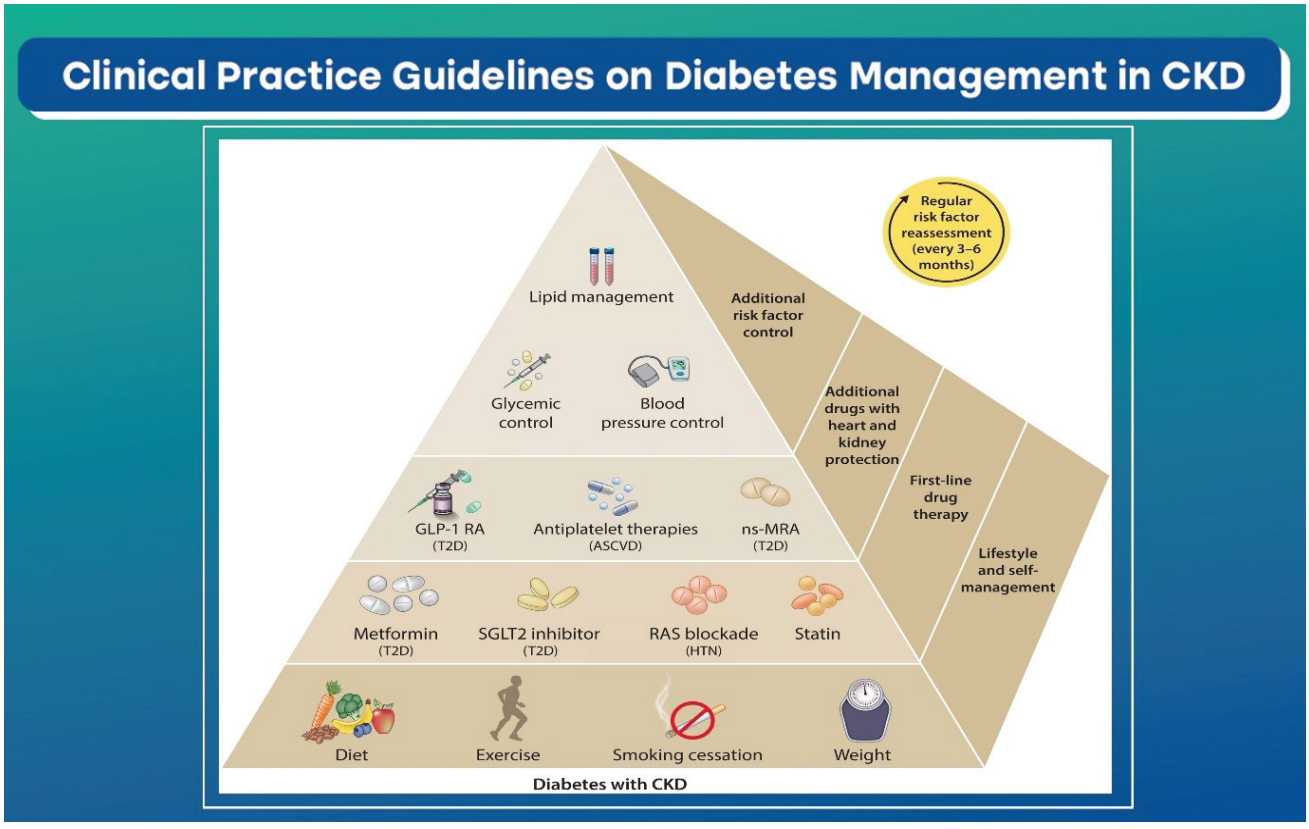

Figure 1: KDIGO 2022 Clinical Practice Guidelines on Diabetes Management in CKD (8)

Glycemic control:

The Diabetes Control and Complications Trial was a pivotal study that demonstrated the benefits of intensive blood glucose control in minimizing the risk of microalbuminuria and impaired kidney function among individuals with type 1 diabetes (5). Participants who received intensive glycemic treatment (mean HbA1c 7%) experienced a 35-45% lower likelihood of developing microalbuminuria compared to those in the control group (mean HbA1c 9%) (5).

Similarly, the United Kingdom Prospective Diabetes Study investigated participants with type 2 diabetes and reported that participants who attained a mean HbA1c level of 7.0% exhibited a 24% reduced risk of developing microalbuminuria after nine years of intensive treatment, compared to those in the standard care group with a mean HbA1c of 7.9% (6). Even though glycemic control decreased over time, those who initially received intensive treatment continued to experience a lower risk of vascular complications and reduced all-cause mortality during the subsequent ten-year follow-up period (6).

The ADA 2022 guidelines recommend a target A1c of 7.0% to slow the progression of microvascular complications of diabetes, including DKD, in patients with both type 1 and 2 diabetes (7).

Blood Pressure Control:

Effective management of hypertension is essential for preventing CKD progression, DR, and cardiovascular complications (4,8). It is widely agreed that for patients with diabetes, hypertension, and albuminuria, initiating treatment with a renin-angiotensin system (RAS) inhibitor - specifically an angiotensin-converting enzyme inhibitor (ACEi) or an angiotensin II receptor blockers (ARB) - is crucial (4,8). These medications should be titrated to the highest tolerated dose (7–9).

Two key studies examining the impact of RAAS blockade on the progression of Diabetic Nephropathy (DN) and DR in patients with T1DM were the Renin-Angiotensin-Aldosterone System Study and the Diabetic Retinopathy Candesartan Trials (10). These studies found that RAAS blockade slowed the advancement of DR but had no effect on the progression of DN over time (10). The Bergamo Nephrologic Diabetes Complications Trial, which compared an ACEi to calcium channel blockers, was a landmark study that demonstrated the beneficial effect of RAAS inhibitors in delaying the progression of DN in patients with T2DM (11). This trial showed less progression to the stage of microalbuminuria with the ACEi compared to the calcium channel blocker (11).

Treatment of Dyslipidemia:

Clinical studies have suggested that lipid-lowering therapies such as statin therapy may be effective in slowing the progression of DN (3). The Action to Control Cardiovascular Risk in Diabetes-Lipid trial and the Diabetes Atherosclerosis Intervention Study both demonstrated a slower progression to microalbuminuria (12). Furthermore, the Diabetes Atherosclerosis Intervention Study found that treatment with fenofibrate significantly reduced the rate of progression from normal albumin levels to microalbuminuria in patients with type 2 diabetes over 3 years compared to placebo (13).

PHARMACOLOGICAL INTERVENTIONS

RAAS Blockade:

The use of RAAS inhibitors (i.e., ARBs and ACEi) has been the mainstay in managing DKD, with evidence showing their effectiveness in slowing DKD progression in patients presenting with albuminuria and hypertension (14). These medications should be titrated to the highest dose the patient can tolerate (8).

Clinical studies

Landmark clinical trials such as the Reduction of Endpoints in Non-Insulin-Dependent Diabetes Mellitus with the Angiotensin II Antagonist Losartan (RENAAL) and Irbesartan in Diabetic Nephropathy trials have shown that ARBs reduced the primary outcomes of doubling of serum creatinine, ESKD, and mortality compared to standard hypertension management (15,16).

Sodium-glucose cotransporter-2 inhibitor (SGLT-2i):

SGLT2 inhibitors are used as antihyperglycemic treatments for type 2 diabetes, acting by increasing urinary glucose elimination through the inhibition of glucose and sodium reabsorption in the proximal renal tubules (14). Strong evidence supports that SGLT2 inhibitors effectively lower the risks of ESKD, cardiovascular mortality, and heart failure hospitalizations (14). These cardiovascular and renal benefits are independent of the antihyperglycemic effect, which diminishes with a lower eGFR (14).

Clinical Evidence

The CREDENCE trial was the first significant placebo-controlled study evaluating an SGLT2 inhibitor, where the primary focus was on renal outcomes (21). The findings demonstrated that canagliflozin in patients with type 2 diabetes and kidney disease resulted in a 30% relative risk reduction of kidney disease compared to placebo (21). Furthermore, the DAPA-CKD trial conducted in 2020 was the first kidney disease outcome trial to include a substantial proportion of participants both with and without type 2 diabetes (22). The study enrolled participants with an eGFR range of 25–75 mL/min/1.73m2 and albuminuria of 200–5,000 mg/g (22).

The results showed that the primary kidney outcome was reduced by 39% in the dapagliflozin group compared to placebo, with similar effects observed in patients with and without type 2 diabetes (22). Importantly, dapagliflozin also demonstrated a 31% reduction in all-cause mortality in both populations (22).

Mineralocorticoid receptor agonists (MRAs):

MRAs represent an important component of RAAS blockade, with multiple studies highlighting their potential benefits in managing hypertension and reducing proteinuria (14). Additionally, MRAs have been shown to decrease albuminuria and mitigate secondary markers of renal fibrosis and inflammation (14). While steroidal MRAs such as spironolactone and eplerenone have been available for some time, their clinical use is somewhat restricted (14). The latest advancement in this class of medications is the development of non-steroidal MRAs (ns-MRAs), with finerenone being the prototype (14). These newer agents have demonstrated protective cardiorenal effects and offer a more favorable risk-to-benefit ratio compared to their steroidal predecessors (14).

Clinical Studies

In Phase III trials such as FIDELIO-DKD and FIGARO-DKD, finerenone has demonstrated significant cardiorenal benefits (17). The FIDELIO-DKD trial reported a composite kidney outcome of 5.5% in the finerenone group compared to 7.1% in the placebo group (17). The FIGARO-DKD trial showed a kidney outcome of 9.5% in the finerenone group versus 10.8% in the placebo group among patients with stage 2 and 4 CKD (17).

Both trials confirmed that finerenone effectively reduces cardiovascular and renal complications observed with maximum-dose ACE inhibitors or ARBs, with a lower incidence of hyperkalemia compared to placebo (17).

Glucagon-like peptide-1 receptor agonist:

Incretin-based therapies, which include dipeptidyl peptidase-4 inhibitors (DPP4i) and glucagon-like peptide-1 receptor agonists (GLP-1 RA), target postprandial neuroendocrine pathways to enhance pancreatic insulin secretion, suppress glucagon release, increase satiety, and delay gastric emptying (14). In addition to their antihyperglycemic effects, these therapies have been shown to improve cardiovascular and renal risk factors, specifically blood pressure, body weight, and lipid profile (14). As a result, this drug class is recommended as the first-line anti-hyperglycemic medication by the ADA, particularly for patients with overweight or obesity, and for those with non-proteinuric kidney disease (7).

Clinical Studies

- Analysis of data from the ELIXA trial, which compared lixisenatide to placebo, demonstrated that lixisenatide reduced the urinary albumin-to-creatinine ratio. This effect was statistically significant only in participants with stage A3 albuminuria (18).
- The LEADER trial, which studied liraglutide versus placebo, revealed renal benefits in patients with type 2 diabetes, with lower rates of nephropathy events observed in the liraglutide group compared to the placebo group (19).
- The SUSTAIN-6 trial, which investigated semaglutide versus placebo, with nephropathy as a secondary outcome, found that new or worsening nephropathy occurred in 3.8% of the semaglutide group versus 6.1% of the placebo group (20).

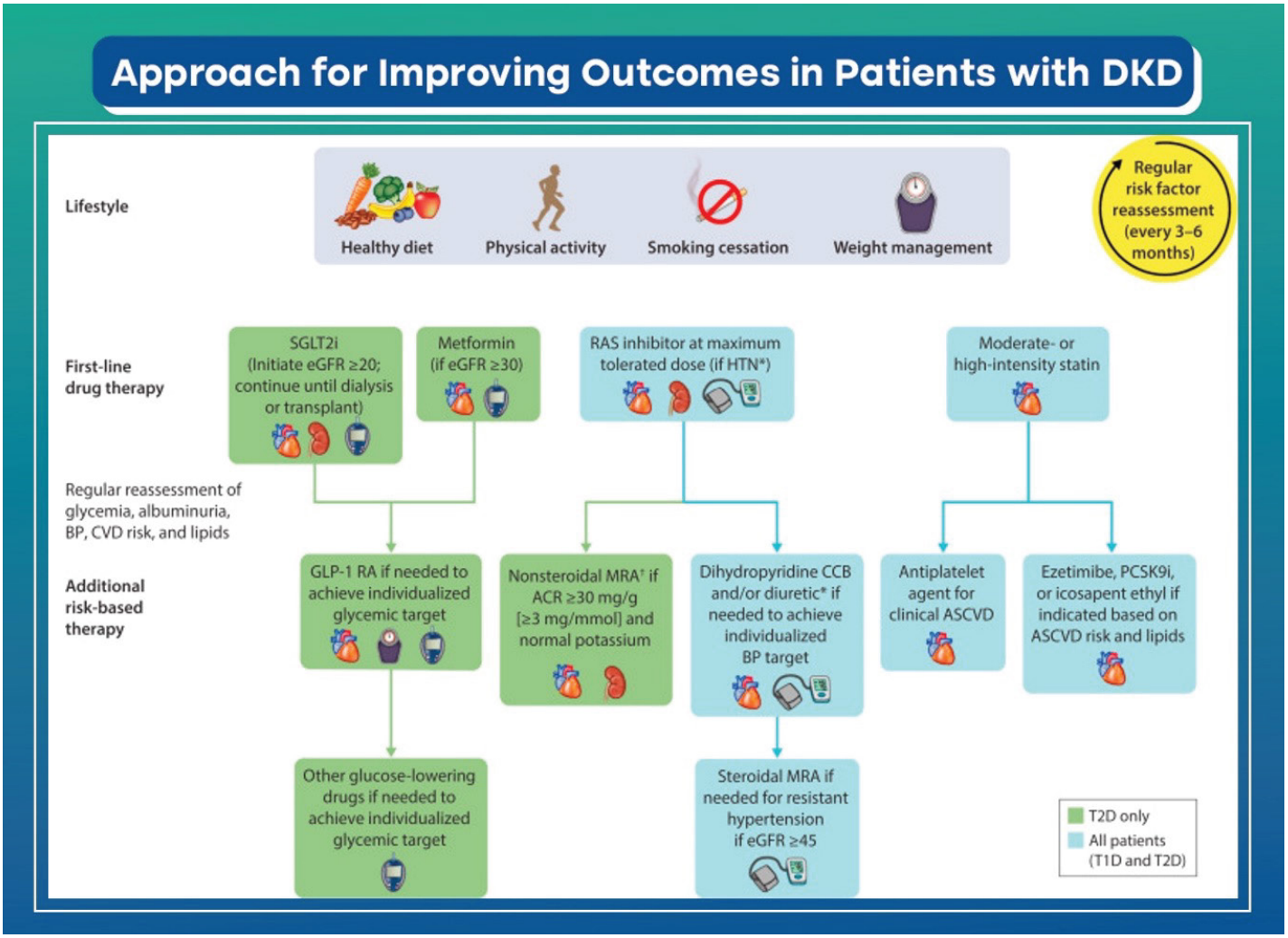

Figure 2: KDIGO 2022 Clinical Practice Guidelines on Diabetes Management in CKD (8)

EMERGING THERAPIES

Novel treatment modalities for DKD include antifibrotic interventions utilizing pirfenidone or pentoxifylline, as well as inhibition of Nox4/1 and chemokine/cytokine pathways (14). Additional pharmacological options targeting various inflammatory cascades have also been the subject of intensive research (14). Most recently, the JAK2/1 inhibitor baricitinib was found to reduce albuminuria in individuals with type 2 diabetes and diabetic kidney disease (14). Moreover, bardoxolone methyl has been investigated for its ability to activate the Keap1/Nrf2 system, which has a crucial role in defense mechanisms against oxidative stress (14).

Key Highlights

- Diabetic kidney disease (DKD) affects 40% of individuals with diabetes, leading to chronic kidney disease and end-stage renal disease globally (1).
- Accurate diagnosis of DKD is based on measuring eGFR and albuminuria, considering clinical features such as diabetes duration and the presence of diabetic retinopathy (2).
- Management of DKD includes lifestyle modifications, pharmacotherapy with RAAS blockade (ACEIs or ARBs), and therapies like SGLT-2 inhibitors, MRAs, and GLP-1 receptor agonists (13).
- Novel treatments for DKD include antifibrotic agents like pirfenidone, JAK1/2 inhibitors such as baricitinib, and the antioxidant bardoxolone methyl, which targets various inflammatory and oxidative stress pathways (13).

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"Rare and loud cause of Heart Failure in a Young Patient" diagnosed and managed successfully at Aster Hospital, Sharjah

Ruptured Sinus of Valsalva Aneurysm (RSOVA) from Non Coronary Sinus (NCS) to Right Atrium (RA) / Venturi Aortic Regurgitation

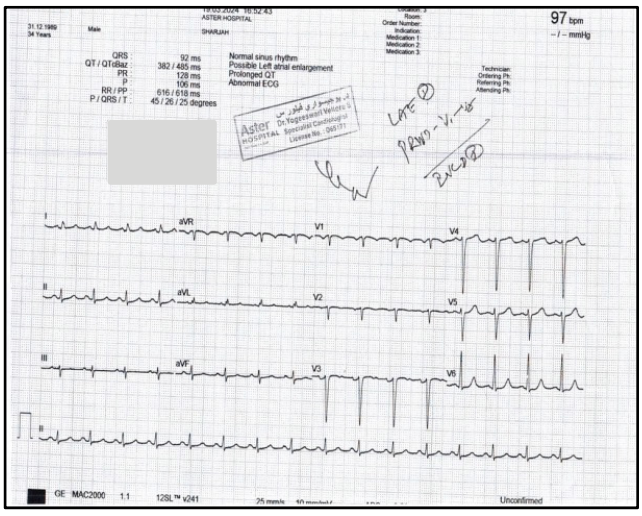
Dr. Yogeeswari Vellore Satyanarayanan
Cardiology (Specialist)

PRESENTATION

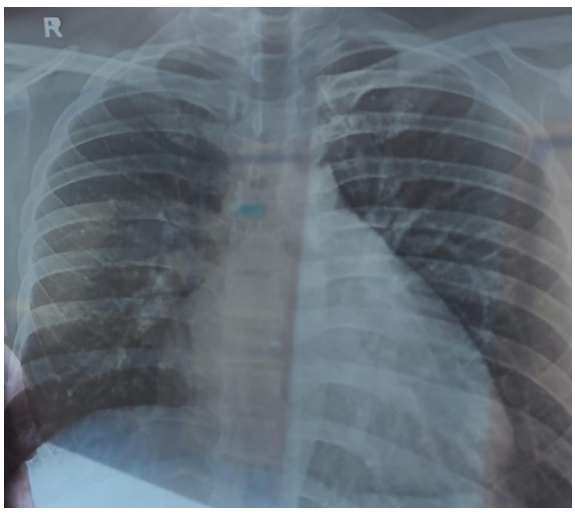
- 34 year old male
- Referred from outside clinic in view of cardiomegaly noted on Chest X-ray with c/o cough for 1½ months
- Referral GP note mentioned multivalvular heart disease in failure that needs open heart surgery
- Complaints of Paroxysmal Nocturnal Dyspnea (PND)
- Occasional yellow-coloured sputum for last 2 days
- LOA, LOW +
- No palpitation/dizziness

CLINICAL EXAMINATION

- BP - 107/49 mmHg, continuously coughing during the examination
- Sats 99% on room air
- No pedal oedema



ECG showing Sinus Tachycardia/ LAE/ Pseudoinfarct Pattern



CXR s/o Cardiomegaly with Pulmonary Venous Congestion (PVC)

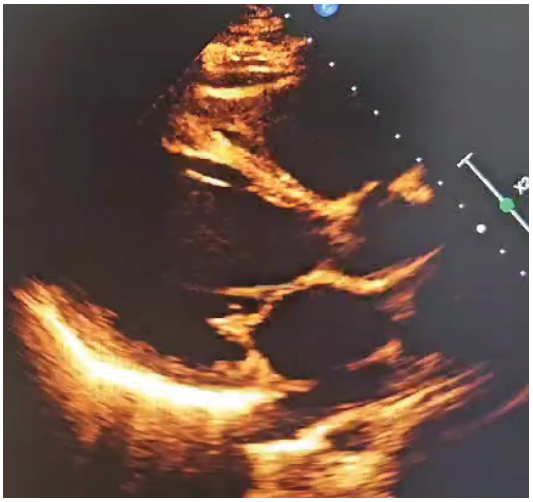
EXAMINATION (Bedside Screening Echo Post-ROSC)

Echocardiogram Findings:

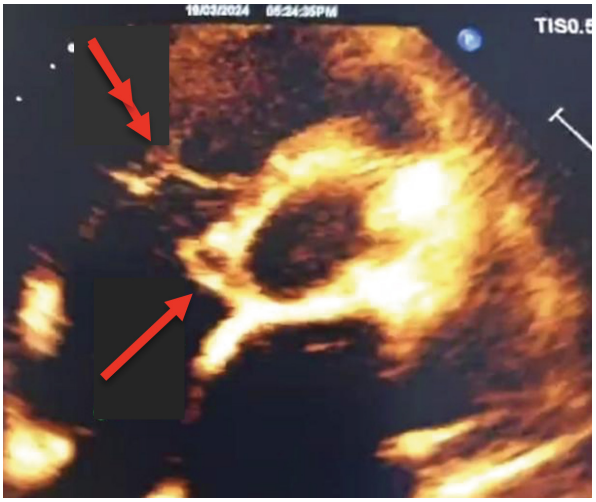
- Dilated LV, adequate LV function (non-brisk LV)
- Moderate AR, no obvious aortic valve pathology
- RSOVA involving NCS - rupturing into RA

Contrast Echo with Agitated Normal Saline showed:

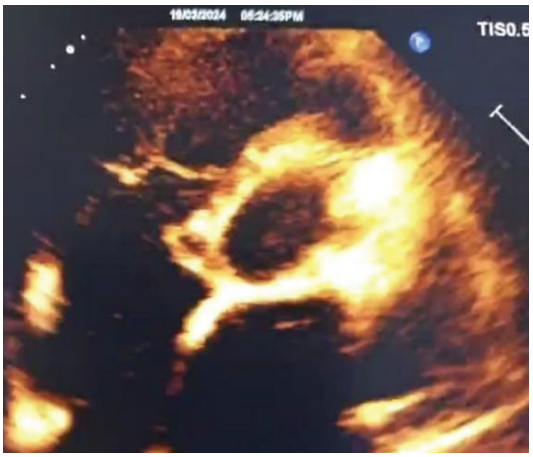
- No evidence of intracardiac/extracardiac intrapulmonary shunts



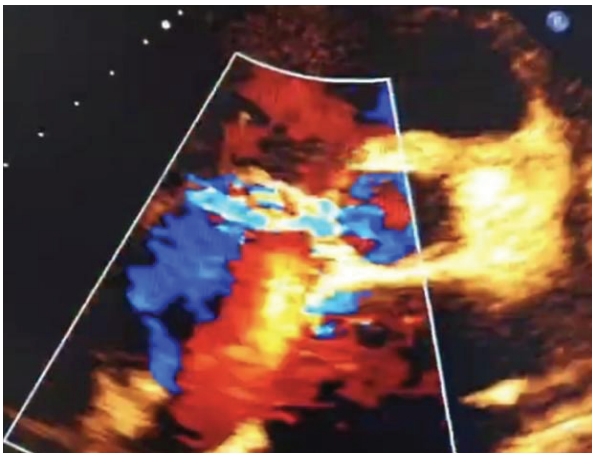
Dilated LV



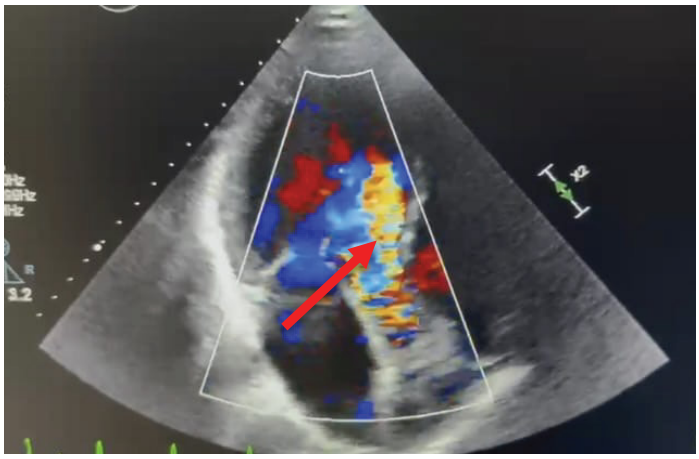
Single arrow - RSOVA into RA
Double arrow - Tricuspid Valve



RSOVA into RA



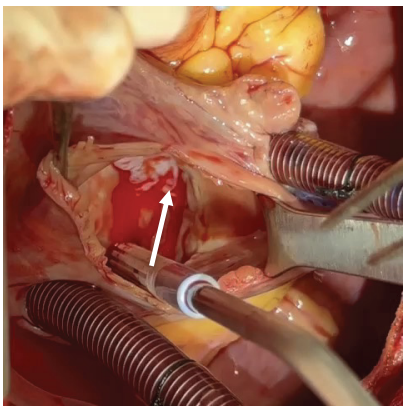
Turbulent Color Doppler signals across RSOVA from NCS to RA



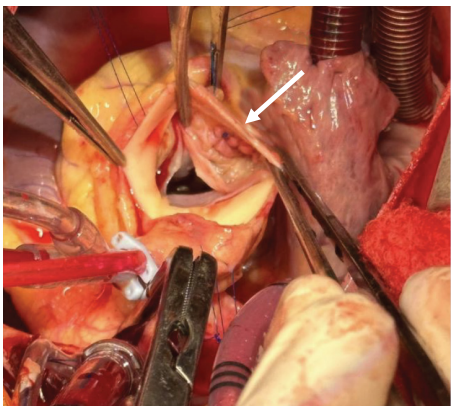
Transesophageal Echo showing AR (Aortic Regurgitation) - no Ventricular Septal Defect (VSD)

IMPRESSION

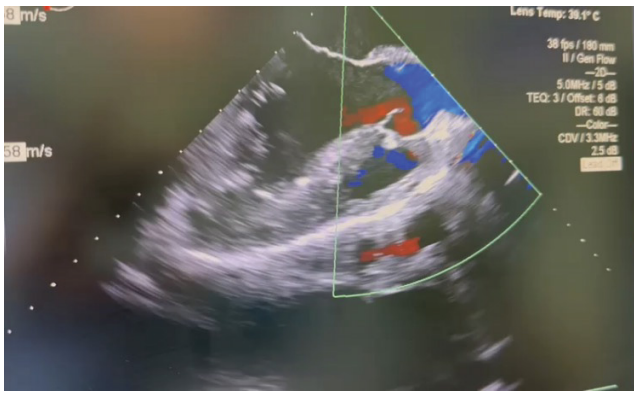
- Young male patient with no known cardiac comorbidities or family history
- Sudden onset of symptoms / Acute Decompensated Heart Failure
- A murmur, loud enough to be picked by GP amongst busy practice
- Cardiomegaly / Dilated LV / AR? Venturi Effect? Valvular
- RSOV aneurysm from NCS (Non-coronary sinus) to RA (Right atrium)



Windsock Deformity



Resection and Pericardial Patch Closure



Post-op TEE - Trivial AR

MANAGEMENT

The patient was shifted to the nearest CTVS centre as he was haemodynamically unstable and underwent resection of the windsock deformity with pericardial patch closure via open heart surgery.

DISCUSSION

RSOVA (Ruptured Sinus Of Valsalva Aneurysm) is an uncommon heart defect that can be congenital or acquired. The surgical prevalence is around 0.2-0.9% (female<male), predominantly affecting males in the Asian population.

The Right coronary sinus is most commonly involved, followed by the Non-coronary sinus and rarely the Left coronary sinus. Once RSOVA is diagnosed, it is necessary to look for associated conditions: Ventricular Septal Defect (VSD), Aortic regurgitation (AR), and Infectious Endocarditis. Haemodynamic stability and associated conditions determine the further management plan: Transcatheter or Surgical Closure.

AR in a patient with RSOVA can be due to either the venturi effect or structural abnormality of the aortic valve leaflets.

Venturi Effect: High-velocity flow through RSOVA results in suctioning or pulling the non-coronary cusp, thereby distorting the anatomy and leading to non-coaptation of Aortic valve leaflets.

In this case, the patient had moderate to severe AR. However, the Aortic valve leaflets were intact during the water test, and hence, the aortic valve was not repaired. Post-op transesophageal echocardiography scans also confirmed the same.

Water Test: This is a simplified test to see if the valve is opposing well 'even with low pressure". The coaptation line and any slack in the leaflet are examined. It helps to decide intraoperatively if the patient needs additional aortic valve repair to correct it then and there instead of re-clamping the aorta and arresting the heart again.

Early intervention (either transcatheter device closure or surgical closure) is a must as it carries high (90%) mortality at the end of 1 year if left untreated.

Multimodality imaging, including transthoracic (TTE), Transesophageal (TEE), CT, Cardiac MRI, and aortography, helps identify associated lesions and decide on the intervention route: Transcatheter or Surgical.

Size of the defect >5 mm, presence of AR, associated VSD, distance of the RSOVA from the coronary origin <7 mm, etiology and haemodynamic stability favour surgical approach. However, TCC (Transcatheter Closure) can also be performed in the above circumstances if the patient is haemodynamically too unstable to be planned for surgical closure.

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