Aster HOSPITALS & CLINICS
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HealthNews DIGEST

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29th Edition

Dr. Sherbaz Bichu

CEO & Specialist Anaesthetist
Aster Hospitals & Clinics, UAE

On behalf of Aster's leadership, I welcome you to the 29th edition of the HealthNews Digest. I want to reflect on the impressive accomplishments and landmarks our doctors have made in the past year as we celebrate the start of a new one. Despite our unprecedented challenges, you persevered to give our patients outstanding care while adjusting to new therapies and technology.

I appreciate all that you do and am proud of every one of you for your unwavering dedication and commitment to the field of medicine. I have no doubt that we will continue to do great things together. Let's keep coming up with new ideas and pushing the limits of healthcare while prioritising our patients' needs and welfare.

Wishing you all a happy and healthy New Year 2025!

Dr. Ramanathan V

Medical Director
Aster Hospitals & Clinics, UAE

As the Medical Director for Aster Hospitals and Clinics, I am delighted to greet all our doctors as we commence the 29th edition of HealthNews. I want to acknowledge our doctors' incredible work in handling critical cases, achieving milestones in the medical field and providing exceptional patient care by taking on challenging cases and putting cutting-edge treatments into practice that have significantly impacted the lives of the people we serve.

Please keep pushing the boundaries of what is possible in healthcare. Let us continue to build on our triumphs as we enter the new year and make even more significant strides in patient care.

Wishing you and your loved ones a happy and healthy New Year 2025!

Dr. Brijesh Puthalonkunath Valsalan
Orthopaedics (Specialist)

Dr. James Chettupuzhakaran George
Orthopaedics (Specialist)

A case of Revision Total Knee Arthroplasty Surgery done successfully at Aster Hospital, Sharjah

PRESENTATION

- 68-year-old female
- Medical history of Diabetes Mellitus, Hypertension and Hyperlipidemia, on medications
- Surgical history of Total Knee Replacement surgery done 9 years back. The patient was fine only for a year, and then the pain started
- No history of trauma
- Admitted with –
  - Severe pain in the right knee for a year

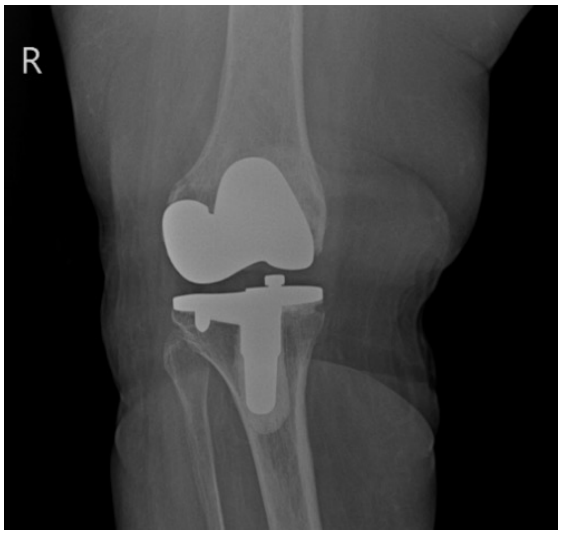
FINDINGS

During Examination:

- The patient was made to stand up and walk; she took two steps with great difficulty
- Antalgic gait
- Previous surgical scar midline on the right knee
- No redness / local warmth
- Genu varum bilateral
- Passive and active ROM 0-90
- Varus stress test - positive, Valgus stress test - negative
- Patellar tracking was normal

Investigations:

- Two synovial fluid studies were performed, and the culture was negative
- X-rays of knees showed a previously operated knee with total knee arthroplasty with malposition and loosening of implants.



Pre-op X-ray Image

DIAGNOSIS

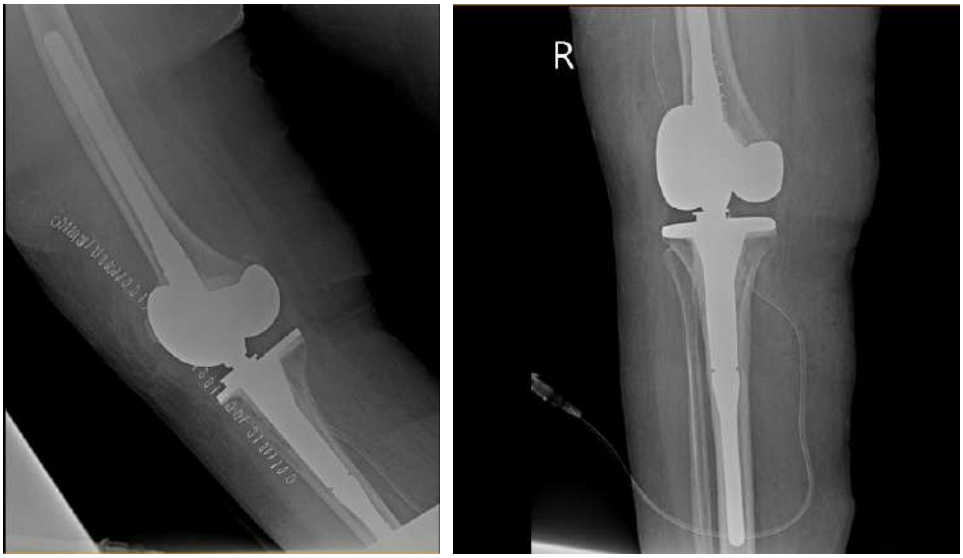
- Painful Total Knee Arthroplasty in the right knee with aseptic loosening

DURING PROCEDURE

The patient was admitted for revision Total Knee Arthroplasty surgery on the right side under spinal and epidural anaesthesia after the surgical site identification:

- The area was prepped and draped in the usual sterile manner.
- The patient was positioned supine. The tourniquet was inflated to 270 mm Hg.
- An incision was made using the midline median parapatellar approach. The joints were exposed in layers.
- Loosening of the prosthesis, bone defects in the femur, deficient LCL, and synovitis were noted.
- Synovial fluid was clear, and the sample was taken for culture. Synovial tissue and samples from the intramedullary canal were taken. Five samples in total were taken for culture and one for histopathology.
- Prosthesis in the femur and tibia was extracted gently using osteotomes at the implant cement interface.
- Femur was loose and came out easily. Then, all the bone cement was taken out taking care to preserve bone as much as possible.
- Significant osteolysis with bone loss was noted in the femur with loss of condyles and AORI Type 2B defect, while the tibia had a Type 1 defect.
- Bone preparation was done in the tibia and femur.
- Intramedullary prep was done with bone cuts for implantation.
- Due to extensive bone defects and poor bone quality, a rotating hinge prosthesis was implanted.
- To make up bone defects, augments were used in the distal medial condylar region and posterior aspect lateral condylar region, and the wound was thoroughly washed using pulse lavage.
- Trial implantation was done and checked for alignment, knee ROM and stability.
- After it was found satisfactory, cementation of tibial and femoral implants was done after washing and drying.
- Zimmer NexGen rotating hinge system was used.
- The excess cement was removed and lavage again once the cement was set.
- Following this knee ROM, alignment and stability were reassessed.

- Tourniquet was released. Hemostasis was achieved. And Gentamicin wash was given.
- Wound was closed in layers over suction drain.
- Compression dressing was done. The dorsalis pedis pulse was checked and found to be normal.



Post-op X-ray Images (Lateral and Anterior-posterior view)

POST PROCEDURE

The postoperative period was uneventful. The patient tolerated the procedure well without any complications. She was closely monitored for 24 hours in the ICU post-op and was then shifted to the ward. Post-op analgesia was given using epidural infusion. Thromboprophylaxis was initiated pre-op and continued throughout the perioperative phase using mechanical measures and Clexane. Drain was removed, the urinary catheter was removed, and the patient was ambulated.

She walked comfortably with walker support and full weight bearing. Physiotherapy continued. She was comfortable and not in pain at the time of discharge.

DISCUSSION

Total Knee Arthroplasty (TKA) has been a revolution for patients with end-stage knee arthritis, resulting in marked improvement in lifestyle and mobility. Although most patients experience substantial symptomatic relief after TKA, up to 20% of patients are unsatisfied with their outcome.

Painful TKA is the biggest cause of this dissatisfaction, and every painful case must be meticulously assessed to offer an appropriate solution.

A useful way of classifying common causes of painful TKA would be to classify as early and late causes:

Early causes:

- Implant instability (related to surgical and technical errors)
- Problems of the extensor mechanism (patella not resurfaced, malalignment of femoral, tibial, or patellar component, tendons failure or degeneration).

Late causes:

- Aseptic loosening
- Infection

Unusual causes of pain are reflex sympathetic dystrophy, synovitis, and hypersensitivity to metal implants are represented.

Practically, the most common causes that need revision include aseptic loosening, infection, and instability. Infection also causes early loosening. A thorough workup to diagnose/rule out infection is vital as infection management differs from aseptic loosening/instability, both of which can be managed by single-stage revision. In contrast, infection would necessitate a two-stage approach with initial debridement and placing of cement spacer and, later on, a second stage revision arthroplasty once all signs of infection have subsided.

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An Overview of Endometrial Hyperplasia

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INTRODUCTION

Endometrial hyperplasia, with or without atypia, is a precancerous condition characterized by a range of morphological changes in the endometrium (1). These changes are marked by an increased gland-to-stroma ratio compared to normal proliferative endometrium (1). The incidence is estimated to range from 133 to 208 cases per 100,000 women, with non-atypical hyperplasia accounting for 121 of these cases (2). Non-atypical endometrial hyperplasia (NEH) carries a 5% risk of progressing to carcinoma, while the risk increases to as much as 30% in atypical hyperplasia (AH) (2). Common symptoms include intermenstrual bleeding, postmenopausal bleeding, menorrhagia, and abnormal uterine bleeding (AUB), particularly in patients undergoing tamoxifen or hormone replacement therapy (2). Risk factors include obesity, hypertension, diabetes, hormone therapy, lack of exercise, and contraceptive use (2). Additionally, postmenopausal, nulliparous, and infertile women are at high risk of EH (3).

Diagnosis of EH involves taking a thorough history, evaluating symptoms such as AUB, conducting radiological examinations like transvaginal ultrasound, CT, or MRI, and performing histological analysis of the endometrial tissue (4). For treatment, conservative approaches like hormone replacement therapy, intrauterine devices, and progestins may be used, while severe cases may require hysterectomy or hysteroscopic resection (4).

This article will provide an overview of endometrial hyperplasia, focusing on the diagnosis and management of EH.

CLASSIFICATION OF EH

According to WHO 2014, EH is differentiated into two categories:

a) **Hyperplasia without atypia** refers to the proliferation of endometrial glands with irregular size and shape but without significant cytological atypia (5). Approximately 1-3% of women with this type of hyperplasia will progress to well-differentiated endometrial carcinoma (5). However, the progression rate is generally low, and most cases can be effectively managed or cured through hormonal therapy, curettage, or a combination of both approaches (6). For diagnosis, a threshold gland-to-stroma ratio of 2:1 is commonly used, although a glandular contribution exceeding 55% (a ratio greater than 1:1) may also be considered compatible with the morphological findings (6).

b) **Atypical Hyperplasia/endometrial intraepithelial neoplasia (EIN)** - EIN is a simultaneous change of epithelial cytology and an increased number of endometrial glands in comparison with the stroma within a

morphologically defined region, distinct from the surrounding endometrium or from entrapped normal glands (5,6). When compared to non-atypical hyperplasia, EIN carries a significantly elevated risk of carcinoma, with approximately one-quarter of EIN patients receiving a carcinoma diagnosis within a year (6). For the diagnosis of EIN, glandular crowding with either cytologic atypia beyond what is expected in proliferative endometrium, or a morphologically distinct subclone of glands not attributable to benign metaplasia, is required (6).

CLINICAL MANIFESTATIONS OF EH

Endometrial hyperplasia most commonly presents with AUB (6). Premenopausal patients typically exhibit changes in menstrual cycle frequency, volume, regularity, and interval, often accompanied by intermenstrual bleeding (6). In contrast, postmenopausal women commonly present with vaginal bleeding after menopause (6).

Routine systemic and gynaecological examinations should be performed as part of the clinical assessment (6). Physical examinations may be unremarkable or may reveal metabolic abnormalities, such as increased weight and features of polycystic ovarian disease (6).

EVALUATION AND DIAGNOSIS OF EH

Imaging Examinations

Transvaginal ultrasonography is the preferred method for evaluating endometrial hyperplasia, while transrectal ultrasound is recommended for VIRGO patients (6,7). Postmenopausal women with an increased endometrial thickness detected by ultrasound face a heightened risk of endometrial hyperplasia and endometrial cancer (6,7). Therefore, postmenopausal women with bleeding and an endometrial thickness exceeding 4 mm should undergo further evaluation (6,7). Women taking tamoxifen should also be closely monitored via ultrasound for changes in endometrial thickness, as excessive endometrial thickening is associated with a greater risk of endometrial lesions (6,7).

Diffusion-weighted magnetic resonance imaging can aid in the identification of invasive carcinomas and may also be used to assess endometrial hyperplasia and other endometrial lesions (6,7).

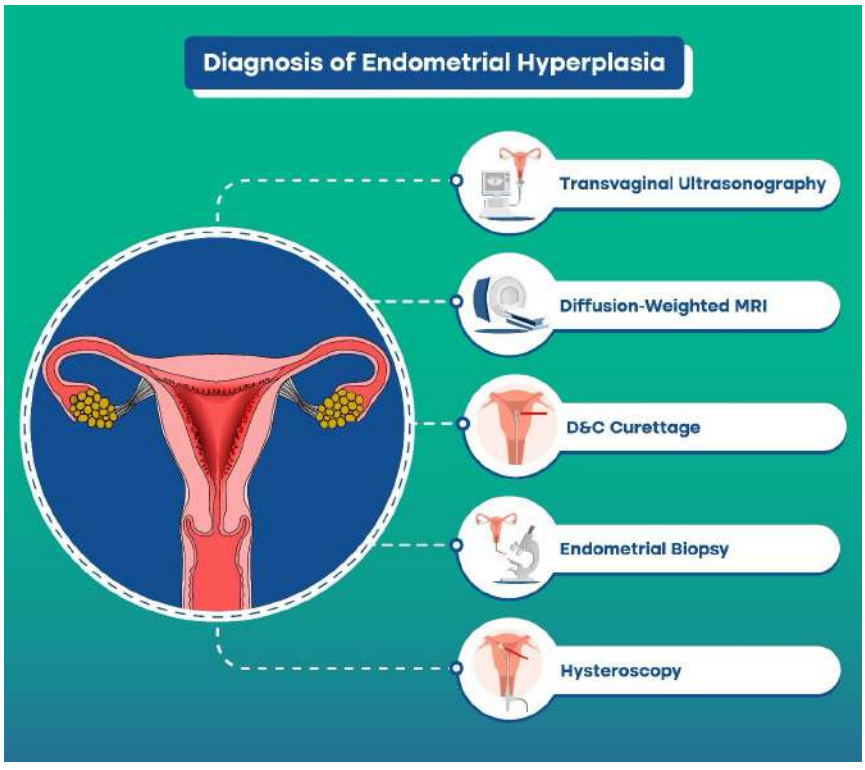


Figure 1: Diagnostic Approaches for EH

Endometrial Biopsy

In all cases of suspected endometrial lesions, a confirmatory endometrial biopsy should be performed (4). Diagnostic curettage and hysteroscopic endometrial biopsy are the traditional biopsy methods utilized (6). Endometrial micro-histopathological examination techniques, such as aspiration biopsy, also serve as highly accurate diagnostic tools (6). Additionally, while not a replacement for histopathological examination, endometrial cytology can be used to screen for endometrial lesions and assist in diagnosis (6).

Hysteroscopy and Hysteroscopic Endometrial Biopsy

Hysteroscopy is a safe and minimally invasive endoscopic procedure that allows for direct visualization and targeted biopsy of the uterine lining (endometrium), as well as assessment of endometrial lesions (6). This technique enables the detection of various endometrial morphological features, including uneven thickening, abnormal vascular patterns, cystic glandular dilation, and structural alterations in the gland duct openings (6).

MANAGEMENT OF EH

Treatment selection is based on patient-specific factors such as age, overall health, presence of cytologic atypia, and fertility aspirations (8). EH without atypia typically responds favorably to progestin-based therapies (8). Hormone therapy is also recommended for individuals whose general health precludes them from tolerating surgery due to coexisting medical conditions (8).

For those with atypical EH or persistent EH without atypia who are symptomatic, hysterectomy is often the preferred management approach (8). For individuals seeking pregnancy, EH treatment presents challenges, requiring conservative management regardless of whether atypia is present (8).

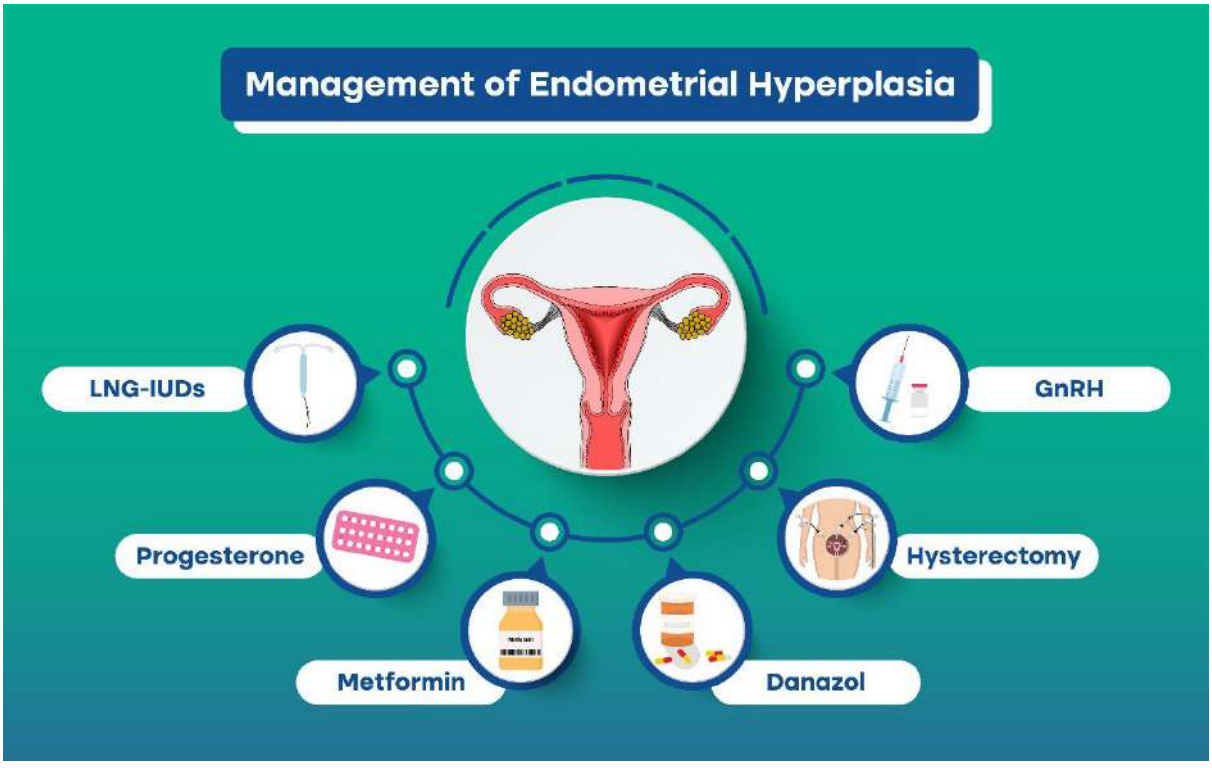


Figure 2: Management Approaches for EH

Progestin therapy

Progestins, synthetic hormones with progesterone-like effects, are commonly used to induce regression of EH in women without atypia or those desiring fertility preservation (8,9). Progestins, either alone or combined with estrogen, offer hormonal contraception and prevent EH caused by unopposed estrogen exposure (8). They reduce glandular cellularity through apoptosis and inhibit angiogenesis in the underlying myometrium in complex EH cases (8,9). Administered orally, intramuscularly, via vaginal creams, or intrauterine devices, progestins effectively reverse EH with or without atypia and reduce EH in 61% of patients with atypical hyperplasia (9).

The mode and duration of progestin treatment are critical (8). EH typically responds within 10 weeks, with significant results after 3 months, and a median resolution time of 6 months (8). If no response occurs, progestin therapy may continue, or hysterectomy may be considered (8).

Medroxyprogesterone Acetate

Medroxyprogesterone acetate (MPA), a synthetic progestogen, is commonly used to manage absent or irregular menstruation and abnormal uterine bleeding (9). In postmenopausal women on estrogen therapy, MPA effectively prevents endometrial overgrowth, lowering the risk of hyperplasia progression (8). Cyclic MPA administration is generally safer and better tolerated than continuous use (8). In a multicenter trial by Ushijima et al., MPA achieved an 82% complete response rate and an 18% partial response rate in endometrial hyperplasia patients, with follow-ups of 25 to 73 months (10). Typical MPA dosing involves 10 mg daily, either continuously for 6 weeks or cyclically for 3 months (9). For partial responders, the dose may be increased to 10 mg four times daily, with treatment extended by 3 months (9).

Levonorgestrel

Levonorgestrel (LNG), a second-generation progestin, is commonly used in hormonal contraceptives and in levonorgestrel intrauterine system (LNG-IUDs) for treating EH (8). It is considered a first-line option for non-atypical endometrial hyperplasia due to its proven effectiveness and favorable safety profile, and it can be maintained for up to 5 years in patients who show a positive treatment response (11).

In a multicenter trial of 170 women with low- or medium-risk EH, LNG-IUD normalized endometrial histology within 6 months and was more effective than cyclic progestogens and continuous oral therapy (12). Among perimenopausal women with non-atypical EH, the LNG-IUD achieved an 88.1% success rate after 12 months (12). A UK study observed 90% histological regression after 2 years, while another study reported 100% EH remission with intrauterine LNG (12).

THERAPIES OTHER THAN PROGESTINS

Danazol

Danazol, a synthetic androgen derivative of 17α-ethinyltestosterone, is commonly utilized as a treatment for endometriosis (8). This compound can induce a hypoestrogenic and hypoandrogenic state within the uterus, leading to endometrial atrophy (8). Multiple studies have demonstrated the significant anti-endometrial hyperplasia effects of danazol (8). Furthermore, it has been proposed as an effective and safe alternative to progestogen therapies for managing endometrial hyperplasia (8).

Danazol-impregnated intrauterine devices have been suggested as a novel and potentially efficacious approach for treating endometrial hyperplasia (8). However, some research has indicated an elevated risk of ovarian cancer among women with endometriosis who receive danazol treatment (8).

Metformin

Metformin, a biguanide class medication, is commonly used to manage type 2 diabetes mellitus and polycystic

ovarian syndrome, particularly among overweight or obese individuals, or in cases where insulin resistance is a significant factor (8). Given the association between insulin resistance and the development of atypical endometrial hyperplasia, as well as the demonstrated anti-proliferative, anti-invasive, and anti-metastatic properties of metformin in various malignancies, the use of metformin represents a logical therapeutic approach for endometrial hyperplasia (8). Interestingly, metformin has been shown to induce the expression of progesterone receptors in endometrial cancer cells, which may enhance the efficacy of progestin therapy or overcome the progestin resistance that can arise due to progesterone receptor depletion during prolonged progestin treatment (8).

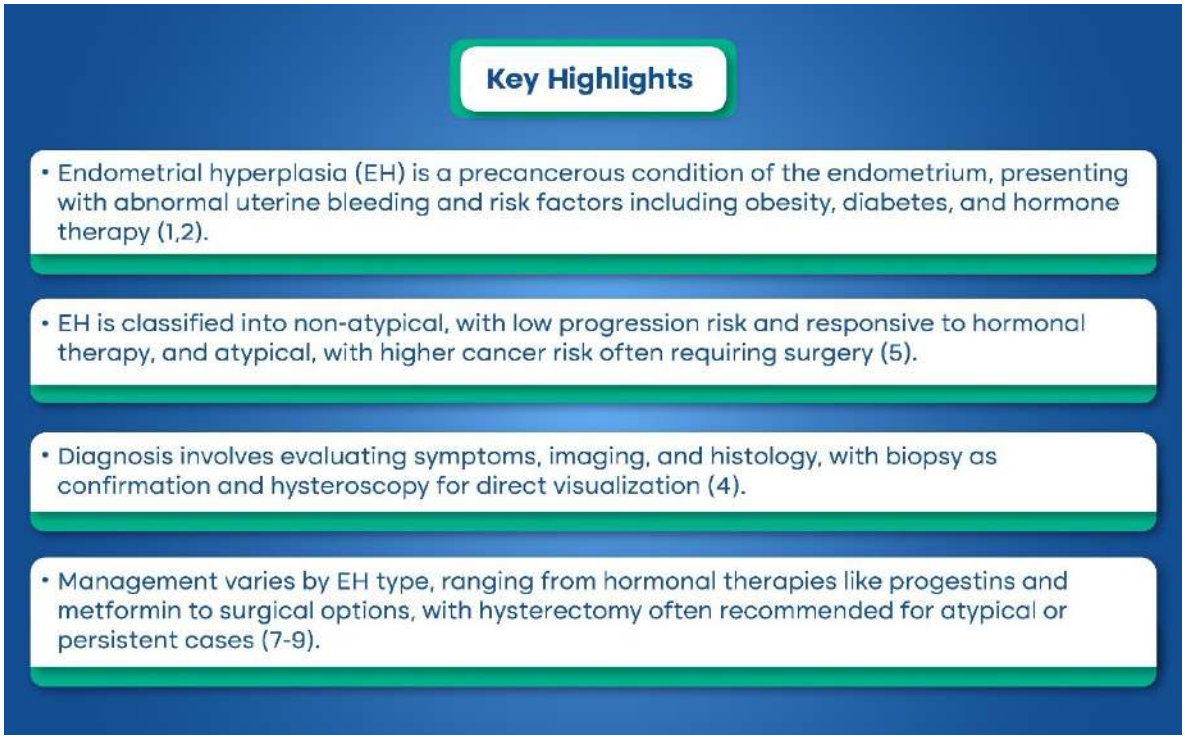
Gonadotropin-Releasing Hormone Therapy (GnRH)

The endometrium contains GnRH receptors, and GnRH agonists can down-regulate these receptors upon prolonged exposure (9). GnRH analogues suppress the hypothalamic-pituitary-ovarian axis, thereby inhibiting estrogen production (9). Consequently, GnRH analogues appear to have a direct anti-proliferative effect on endometrial cells (9). This has led to promising new avenues for the treatment of endometrial hyperplasia (9). GnRH has been administered at a dose of 1 ampule/3.75 mg intramuscularly every 28 days for 6 months to treat women with EH, with or without atypia (9). However, 25% of patients experienced a recurrence of hyperplasia within 16 months of completing therapy (9). A study in which GnRH and tibolone were used to treat EH achieved complete remission in all patients, but with a 19% recurrence rate within 2 years after cessation of therapy (9).

SURGICAL MODALITIES

Endometrial hyperplasia carries the risk of progressing to endometrial carcinoma, prompting a surgical approach as the preferred management strategy for most women with complex EH exhibiting atypia, particularly those who have completed childbearing, do not desire fertility preservation, or have not responded to hormonal therapy (8).

Various surgical interventions have been widely reported as common treatment modalities for atypical EH, such as thermal balloon ablation, laser therapy, or resectoscopic surgery (8). Thermal balloon endometrial ablation or resectoscopic endometrial ablation therapy represents a feasible, safe, and effective treatment option for simple and complex non-atypical EH; however, hysterectomy may be considered a first-line treatment for EH (8). Resectoscopic surgery has demonstrated efficacy in the management of EH without atypia, especially for individuals at high risk for medical therapy or hysterectomy (8). For postmenopausal women with atypical EH, hysterectomy with concurrent bilateral salpingo-oophorectomy is recommended over hysterectomy alone (8).



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A Rare Case of Diabetic Ketoacidosis presenting with Severe Hypertriglyceridemia treated successfully at Aster Hospital, Muhaisnah

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Critical Care Medicine (Specialist)

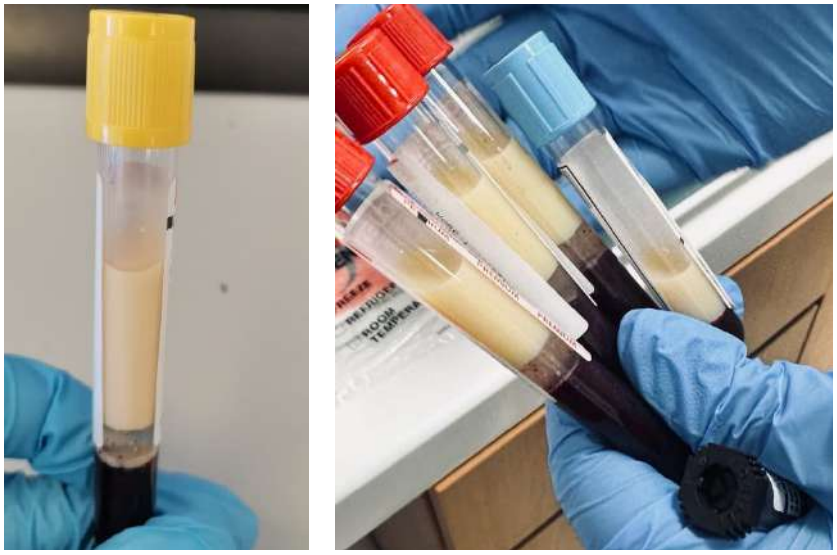
PRESENTATION

- 40-year-old male, African race, 128 kgs
- Bouncer by occupation
- Known case of Diabetes Mellitus and Hypertension, on irregular treatment for 5 months
- Admitted with complaints of:
  - Dizziness and constipation for 3 days
  - Generalized body weakness and tiredness for a day
  - Dryness of mouth

FINDINGS

During Examination:

- Conscious/oriented/afebrile
- Vitally stable
- Heart s1s2 heard, lungs clear
- P/a soft, non-tender
- No evidence of pallor/icterus
- GRBS high (more than the upper limit of glucometer)
- Pupils 2 mm bilateral equal reacting to light
- No focal neurological deficit
- Started on IVF and insulin and shifted to ICU for further management



Lipemic Blood Samples

DIAGNOSIS

Diabetic Ketoacidosis (DKA) presenting with Severe Hypertriglyceridemia.

COURSE IN HOSPITAL

- DKA was managed according to ADA protocol with IV fluids and insulin infusion.
- During the course of treatment, his blood samples collected were lipemic, and triglycerides were more than the upper limit of the lab (>6000). Hence, he was started on high-dose insulin (0.1-03 units/kg/hr) with dextrose infusion to manage his triglyceride levels, and they gradually reduced to 880.
- Regular and intensive monitoring of sugars and electrolytes was done.
- Internal medicine opinion was taken for Diabetes Mellitus control and Hypertension.

POST PROCEDURE

The patient was discharged with stable vitals and asked to follow up with Internal medicine for Diabetes Mellitus, Hypertension and Lipid control. Also, advised for diet modification, daily exercise and avoiding high fat and oily food.

DISCUSSION

Hypertriglyceridemia (HTG) is defined as triglyceride levels of >150 mg/dL (>1.7 mmol/L) and further classified into moderate, moderately severe and severe HTG with defining levels of 150-499,500-999 and greater than 1000 mg/dL respectively. The aetiology of HTG is multifactorial, including insulin resistance disorders, certain medications, estrogens and bile sequestrants, and renal disease. However, severe HTG is mainly attributed to genetic causes such as familial chylomicronemia or type V hyperlipoproteinemia.

Clinical manifestations of HTG vary based on its severity and aetiology. Xanthomas and xanthelasmas are commonly seen in patients with hereditary disorders. Pancreatitis is another common presentation, with the risk of development between 10% and 20% in those with triglyceride levels of >2000 mg/dL. In cases where DKA has induced HTG, triglyceride levels usually do not exceed 1500 mg/dL. There are two reported cases where triglyceride levels exceed 10,000 mg/dL.

In this case, the high protein diet of our patient (90 eggs/day and 3 whole grilled chickens/day) coupled with the fact that he was unaware of his diabetic status, might have caused the rise in triglyceride levels to >6600mg/dL.

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Hypertrophic Cardiomyopathy: Clinical Update on Diagnosis and Management

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INTRODUCTION

Hypertrophic Cardiomyopathy (HCM) is a genetic condition affecting cardiac myocytes, characterized by left ventricular hypertrophy unexplained by secondary causes and a non-dilated left ventricle, with normal or increased ejection fraction (1). It is usually asymmetrical, with the basal interventricular septum exhibiting the most severe hypertrophy (1). Occasionally hypertrophy is restricted to other myocardial regions, such as the apex, the mid-portion as well as the posterior wall of the left ventricle (1). Typically, myocyte hypertrophy, disarray, and myocardial fibrosis are present histologically (1). Prevalence of HCM has been estimated to be 1 in 500 in the general population(1). Globally, HCM affects approximately 20 million people (2). While spontaneous mutations contribute to its burden, most cases arise from inherited autosomal dominant mutations in genes encoding sarcomere or Z-disk proteins, with beta-myosin heavy chain (MYH7) and myosin-binding protein C (MYBPC3) being the most commonly implicated (1). HCM leads to abnormal thickening of the left ventricular walls, which may stiffen over time, reducing the heart's ability to effectively fill and pump blood (3). Symptoms include chest pain, shortness of breath, fatigue, dizziness, fainting, palpitations and peripheral swelling (4). HCM is progressively regarded as a manageable disease, with modern interventions reducing mortality to below 1% and significantly improving patient outcomes (3). However, sudden cardiac death (SCD) remains a concern, especially in individuals under 30, while heart failure remains the leading cause of mortality (5).

This article will give an overview on HCM, covering its pathophysiology, clinical diagnosis, risk factors, and management strategies, including both pharmacological and surgical treatments.

DEFINITION OF HCM

Diagnosis of HCM in adult patients can be established by imaging typically with 2D echocardiography or cardiovascular magnetic resonance (CMR) showing a maximal end-diastolic wall thickness of ≥15 mm anywhere in the left ventricle, in the absence of another cause of hypertrophy in adults (6). Hypertrophy (13-14 mm) can be diagnostic when present in family members of a patient with HCM or associated with a positive genetic test identifying a pathogenic or likely pathogenic variant often in a sarcomere gene (7).

NATURAL HISTORY OF HCM

Even though HCM can be compatible with normal life expectancy without limiting symptoms or the need for major treatments in most patients, many patients can experience significant consequences that are

attributable to the disease (8). Many will experience adverse events, including: (a) sudden cardiac death events; (b) progressive limiting symptoms because of LV Outflow Tract Obstruction (LVOTO) or diastolic dysfunction; (c) HF symptoms associated with systolic dysfunction; (d) AF with risk of thromboembolic stroke and (e) Ventricular arrhythmia (8). Among patients with HCM, cardiometabolic risk factors (e.g. obesity, hypertension, diabetes, obstructive sleep apnoea) are highly prevalent and are associated with poorer prognosis, highlighting the importance of intensive risk factor modification of traditional risk factors (8).

PATHOPHYSIOLOGY OF HCM

HCM is an inherited cardiac condition marked by the presence of unexplained left ventricular hypertrophy (4). It is classified into obstructive and non-obstructive forms, with varying levels of asymmetric left ventricular hypertrophy (9). Obstruction is considered present if peak LVOT gradient is ≥30 mm Hg (10).

The severity of obstruction and clinical manifestations are influenced by the extent of hypertrophy (6). Pathophysiology of HCM consists of dynamic LVOTO, mitral regurgitation (MR), diastolic dysfunction, myocardial ischemia, arrhythmias, metabolic and energetic abnormalities, and potentially autonomic dysfunction (10).

Dynamic LV outflow tract obstruction in HCM arises from the systolic anterior motion (SAM) of the anterior mitral valve leaflet (9). This obstruction is dynamic, resulting from a pressure gradient that pulls the anterior mitral leaflet forward, exacerbating the obstruction (9). LVOTO in HCM is dynamic and sensitive to ventricular preload, afterload and contractility (10). Hence, gradients vary with heart rate, blood pressure, volume status, activity, medications, food, and alcohol intake (10). In about 25% of patients, the obstruction is present at rest, while in others, it can be induced by provocative maneuvers (9). Maneuvers include standing, Valsalva strain, or exercise with simultaneous auscultation or echocardiography (11).

Most individuals with HCM exhibit abnormal diastolic function, leading to increased left ventricular pressures and impaired ventricular filling, which further aggravates obstruction (9). Altered ventricular load with high intracavitary pressures, impaired LV compliance from hypertrophy and fibrosis, altered energetics, microvascular ischemia, and delayed inactivation from abnormal intracellular calcium reuptake are features of HCM that contribute to diastolic dysfunction (7).

As coronary arteries fill during diastole, the combination of ventricular stiffness, outflow tract obstruction, and impaired diastolic filling heightens the risk of myocardial ischemia (9). This ischemia may contribute to ventricular arrhythmias and sudden cardiac death (9). In severe cases, these events can occur at rest but are more commonly triggered by activities such as exercise, which increase myocardial demand (9).

Abnormalities of the mitral valve in patients with HCM include excessive leaflet length, anomalous papillary muscle insertion, and anteriorly displaced papillary muscles (12). MR caused by SAM is usually mid-to-late systolic in timing and posterior or laterally directed jet (12). Patients with HCM may have autonomic dysfunction, with impaired heart rate recovery and inappropriate vasodilatation (12).

CLINICAL DIAGNOSIS OF HCM

Clinical evaluation for HCM usually triggered by the identification of a family history of HCM; by symptoms in response to exertion-chest pain, dyspnea, palpitations, and syncope-including a cardiac event; by detection of a heart murmur during physical examination; during an echocardiographic examination performed for other indications; or by abnormal results on a 12-lead ECG (6). Classically, patients with HCM have a harsh crescendo-decrescendo systolic murmur best heard at left sternal border which intensifies with maneuvers like the Valsalva maneuver that reduce left ventricular volume, prominent apical point of maximal impulse, abnormal carotid pulse, and a fourth heart sound (6).

Figure 1 illustrates the recommended approach for initial diagnosis and the testing algorithm for patients with confirmed or suspected HCM (13).

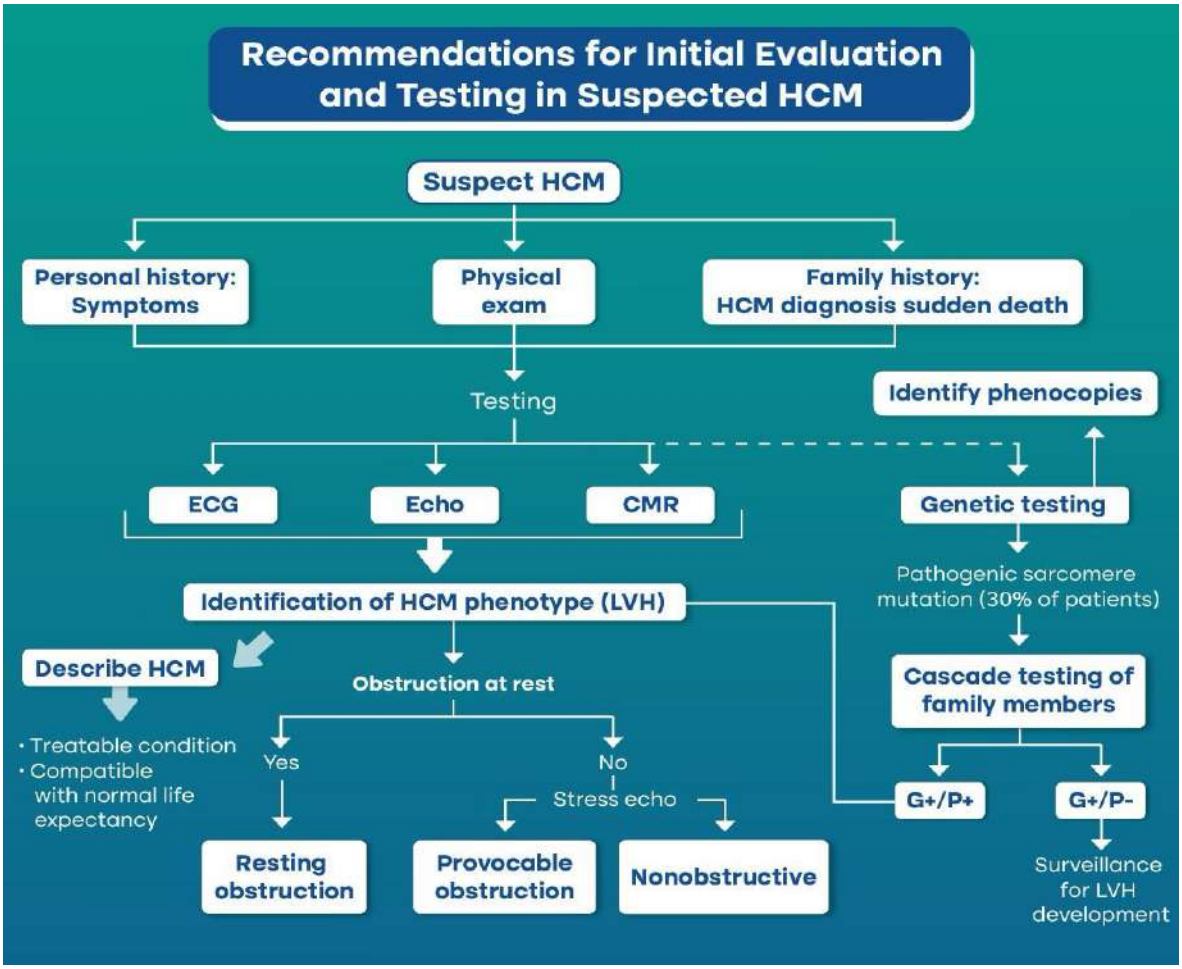


Figure 1: Diagnostic Recommendations for Suspected Hypertrophic Cardiomyopathy (11)

(Abbreviations: CMR - cardiac magnetic resonance; G - genotype; ECG - electrocardiography; echo - echocardiography; HCM - hypertrophic cardiomyopathy; LVH - left ventricular hypertrophy; P - phenotype; SAM - systolic anterior motion)

1. DIAGNOSIS OF HCM WITH LVOTO

HCM with or without LVOTO is diagnosed as per the echocardiographic criteria mentioned above (6). Comprehensive 2D echocardiography has a primary role in establishing the diagnosis of HCM, determining hypertrophy pattern, presence of LV apical aneurysms, LV systolic and diastolic function, mitral valve function, and presence and severity of LVOTO (6).

2. DIAGNOSTIC IMAGING AND TESTS

- **ECG:** It is used to identify electrical abnormalities, including left ventricular hypertrophy, atrial enlargement, and arrhythmias (6).
- **Echocardiography:** The primary tool for diagnosing HCM, echocardiography provides detailed structural and functional imaging to assess left ventricular hypertrophy, SAM, valvular abnormalities , LVOTO and LV function (7).
- **Cardiac MRI:** It provides high-spatial resolution and tomographic imaging to identify myocardial fibrosis (LGE) and structural abnormalities and provide accurate LV wall thickness measurements, quantification of LV and RV chamber size, LV mass, systolic function, and apical aneurysms (7). CMR imaging is therefore a complementary imaging technique in the evaluation of HCM patients for diagnosis, risk prediction, and preprocedural planning for Septal Reduction Therapy (7).

- **Genetic Testing:** It is recommended for patients and their families due to the genetic basis of HCM (7). Detecting pathogenic mutations not only confirms the diagnosis but also guides family screening (7). Pathogenic mutations are identifiable in approximately 50% of patients (7). First-degree relatives should undergo genetic testing and regular cardiac evaluations to monitor for early signs of HCM (7).
- **Cardiac CT:** is considered in adult patients with suspected HCM if the Echocardiogram is not diagnostic and CMR imaging is unavailable (13).
- **Cardiac Catheterization:** For symptomatic HCM patients for whom there is uncertainty regarding presence or severity of LVOTO on non-invasive imaging studies (13).

3. IDENTIFYING PATIENTS AT RISK OF SUDDEN CARDIAC DEATH

Identifying individuals at risk of SCD is a critical aspect of managing HCM (6). Key risk factors include a family history of SCD, unexplained fainting episodes, non-sustained ventricular tachycardia, severe left ventricular hypertrophy (wall thickness ≥30 mm), abnormal blood pressure responses to exercise, overt systolic dysfunction, LV apical aneurysm and extensive intramyocardial scarring (6). For those at high risk, implantable cardioverter-defibrillators (ICDs) are highly recommended as a preventive measure against SCD (6).

MANAGEMENT OF HCM

The management of HCM encompasses symptom relief, risk assessment for sudden cardiac death, lifestyle modifications, pharmacologic treatments, and surgical or catheter-based interventions, with personalized monitoring (2). A more detailed management algorithm is depicted in Figure 2 (2).

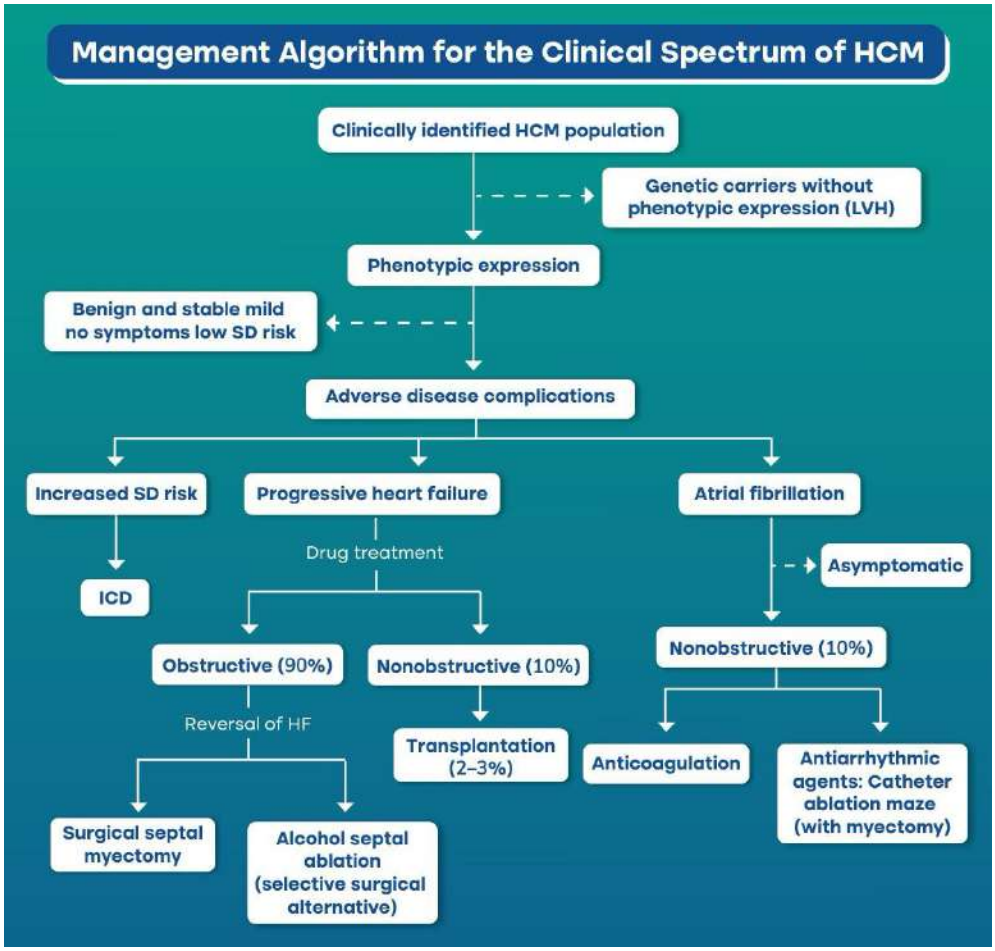


Figure 2: Management of HCM (2)

(Abbreviations: HCM - hypertrophic cardiomyopathy, HF - heart failure, SD - sudden death, ICD - implantable cardioverter-defibrillators)

1. MANAGEMENT OF HCM WITH LVOTO

The management of HCM with LVOTO involves a tailored approach based on symptom severity and individual risk factors (2).

- **Asymptomatic Patients:** In asymptomatic individuals with HCM and LVOTO, regular follow-up is essential (2). Periodic echocardiography is recommended to monitor the degree of hypertrophy and obstruction (2). Routine clinical evaluations are conducted to identify emerging symptoms or complications early (2). Patients are advised to adopt lifestyle modifications, including avoidance of intense physical activities and maintaining adequate hydration, to mitigate the risk of adverse cardiac events (2).
- **Symptomatic Patients:** For symptomatic patients, a combination of pharmacological and invasive approaches is often required for optimal management (14).

A. PHARMACOLOGICAL TREATMENTS:

- **Beta-Blockers:** Beta-blockers, including metoprolol, atenolol, and propranolol play a crucial role in treating cardiovascular disorders (14). They function by inhibiting the effects of adrenaline, which helps decrease heart rate and myocardial contractility, thereby lowering the oxygen demand of the heart and preventing arrhythmias (14). This mechanism aids in relieving symptoms and reducing the outflow gradient in HCM (14).
- **Calcium-Channel Blockers:** Calcium-channel blockers such as verapamil and diltiazem are effective alternatives when beta-blockers are not appropriate (14). These medications enhance diastolic function by relaxing the heart muscle, promoting better filling during diastole (14).
- **Disopyramide:** It is a type 1a antiarrhythmic, a sodium channel blocker, commonly prescribed with beta-blockers or calcium-channel blockers (14). Its negative inotropic effects decrease the force of the heart's contractions, which helps lower the outflow tract gradient (14). This action can relieve symptoms like chest pain and breathlessness (14).
- **Cardiac myosin ATPase inhibitors:** Emerging therapies like Mavacamten and Aficamten, aim to reduce contractility in HCM and improve myocardial energetics (14). These medications work by blocking myosin ATPase activity, which lowers the force of heart contractions and targets the root cause of the condition (14). By alleviating LVOTO, they help improve symptoms and enhance overall heart function (14). Mavacamten has been shown to improve LVOT gradients, symptoms, and functional capacity in 30% to 60% of patients with obstructive HCM (15). Mavacamten was well tolerated and has a good safety profile; only a small subset of patients developed transient LV systolic dysfunction, which resolved after temporary discontinuation of the drug (15). In those who develop LVEF <50%, interruption with resumption at lower dose (if LVEF improves) or discontinuation (if LVEF does not improve to >50%) of cardiac myosin inhibitors is required regardless of associated signs and symptoms (16).

B. THERAPIES TO AVOID:

Drugs that cause peripheral vasodilatation , intravascular volume depletion or increasing myocardial contractility thereby increasing LVOT obstruction may be avoided (14).

- **Vasodilators:** These can exacerbate LVOTO by decreasing preload and afterload, increasing obstruction severity (14), e.g. ACE inhibitors, Angiotensin II receptor blockers, Nitroglycerin (14).

- **High-dose Diuretics:** Excessive intravascular volume reduction can worsen obstruction and should be avoided (14), e.g. Furosemide.
- **Digoxin:** increase inotropy which may exacerbate heart symptoms in HCM patients (17).

C. INVASIVE TREATMENTS:

Symptomatic patients unresponsive to medical therapy may require invasive interventions (18). Septal Reduction Therapy with either surgical septal myectomy or alcohol septal ablation (18).

- **Septal Myectomy:** It is considered the definitive treatment for severe LVOTO, in which a rectangular trough that extends distally to beyond the point of the mitral leaflet-septal contact is created in the basal septum below the aortic valve excising a portion of the hypertrophied septum to relieve obstruction and hence abolishes or substantially reduces LV outflow tract gradients in over 90% of cases, reduces systolic anterior motion-related mitral regurgitation, and improves exercise capacity and symptoms (19). Pre-operative determinants of a good long-term outcome are age <50 years, left atrial size <46 mm, absence of AF, and male sex (19).

  In patients with intrinsic/primary mitral valve disease or marked mitral leaflet elongation and/or moderate-to-severe mitral regurgitation, septal myectomy can be combined with mitral valve repair or replacement (20). In patients with AF, concomitant ablation using the Cox–Maze procedure can also be performed (20).

  The main surgical complications are AV nodal block, left bundle branch block (LBBB), ventricular septal defect, and aortic regurgitation, but these are uncommon in experienced centres (except LBBB) (20).

- **Alcohol Septal Ablation:** This minimally invasive technique involves targeted alcohol injection into the septal perforator artery, inducing controlled myocardial infarction to reduce septal thickness (14). This provides gradient reduction, symptomatic relief and enhances quality of life. The complications which may develop are heart block requiring pacemaker insertion and larger residual LV outflow tract gradients while the procedural mortality is lower than isolated myectomy (14). This procedure is less effective if LVOT gradient is more than or equal to 100mmHg and septal thickness more than or equal to 30 mm (21).

2. MANAGEMENT OF HCM PATIENTS WITH ARRHYTHMIA

Individuals with HCM are at an increased risk of arrhythmias such as atrial fibrillation (AF) and ventricular tachycardia (VT), which may result in severe complications like stroke and SCD (22). Management of AF often involves rate control using beta-blockers (e.g. metoprolol, atenolol) or calcium channel blockers (e.g. verapamil, diltiazem) (22). For maintaining sinus rhythm, rhythm control strategies include antiarrhythmic agents like amiodarone, sotalol, or dofetilide, particularly in cases of persistent AF (22). Catheter ablation is considered for patients whose AF is not well controlled with medications (22). Given the high risk of thromboembolism, anticoagulant therapy with medications such as Warfarin, Dabigatran, Rivaroxaban, or Apixaban is essential for stroke prevention (22). In the management of VT, antiarrhythmic drugs such as Amiodarone and Mexiletine are frequently used, with Sotalol as another option (22). For patients at elevated risk of SCD, ICDs are advised (12). These devices play a vital role in monitoring and correcting life-threatening arrhythmias, providing essential protection for high-risk individuals (22).

3. MANAGEMENT OF HCM WITH HEART FAILURE

Heart failure in HCM results from diastolic dysfunction, LVOTO, or a combination of both. Management starts with a detailed assessment, including symptom evaluation, functional status, and echocardiographic studies (6). Pharmacological treatments with beta-blockers such as metoprolol, atenolol, and propranolol are frequently used to reduce heart rate and myocardial oxygen consumption (6). Calcium channel blockers like verapamil and diltiazem improve diastolic function and provide symptomatic relief, while diuretics are carefully administered to manage fluid overload without aggravating LVOTO (6). Recently, myosin inhibitors like Mavacamten and Aficamten have emerged as innovative therapies to address hypercontractility and enhance cardiac performance (6). In cases where symptoms persist despite medical treatment, advanced options such as septal myectomy or alcohol septal ablation may be considered (6).Patients with LVEF<50% and LBBB, Cardiac resynchronization therapy also may be considered (6). Regular follow-ups, including clinical assessments and echocardiography, are critical to adjust treatment strategies and ensure effective long-term care (6).

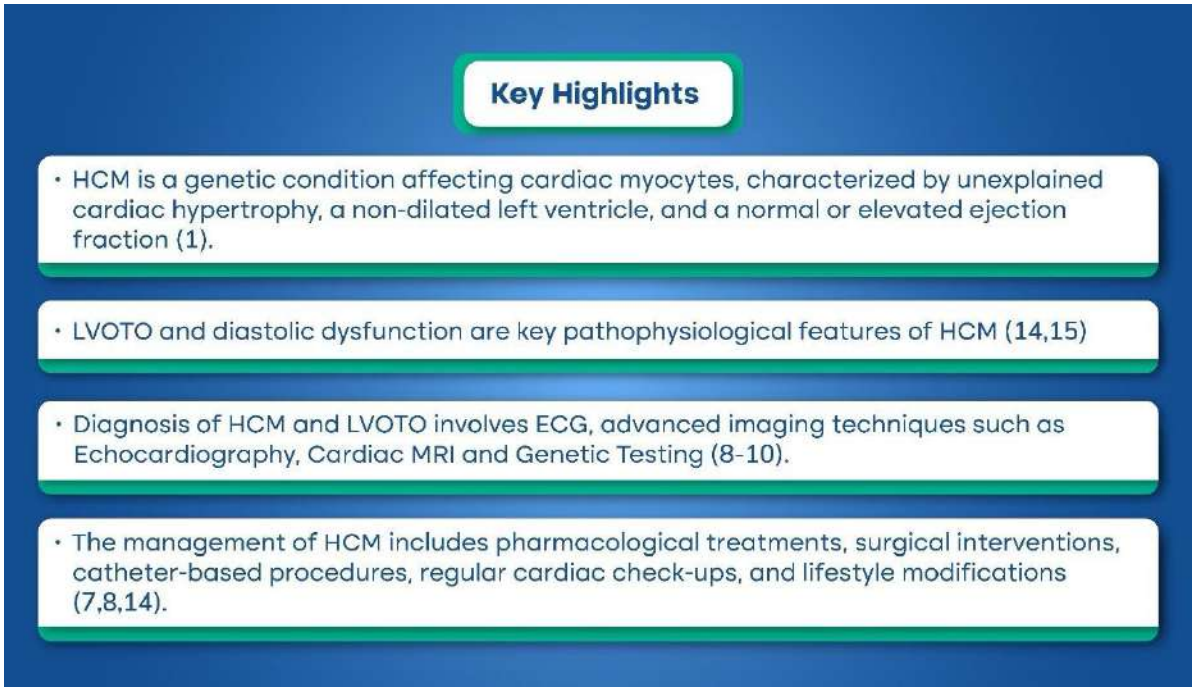
4. LONG-TERM MANAGEMENT AND LIFESTYLE MODIFICATIONS IN HCM

A) ONGOING MONITORING AND FOLLOW-UP

- **Echocardiography:** To monitor structural changes in the heart, such as the degree of LVOTO and the development of hypertrophy, periodic imaging is necessary (7). These assessments provide quick modifications to treatment plans (7).
- **ECG:** Arrhythmias and other potential electrical problems can be detected with regular ECGs and, if necessary, ambulatory ECG monitoring (Holter) to be done (6).
- **Arrhythmia Surveillance:** Continuous vigilance for conditions like AF and ventricular tachycardia is essential, given their potential to cause serious complications such as stroke and sudden cardiac arrest (23).
- **Family Screening and Genetic Advice:** Genetic testing and routine cardiac evaluations are recommended for first-degree relatives to identify early signs of HCM (7). Genetic counselling provides clarity on test outcomes and emphasizes the importance of regular monitoring (7).

B) LIFESTYLE MODIFICATIONS

- **Physical Activity:** Moderate exercise, including activities like walking and swimming, is recommended, while high-intensity or competitive sports should be avoided to reduce the risk of cardiac events (4). Exercise plans should be tailored to align with individual health status (4).
- **Dietary Recommendations:** A balanced diet rich in fruits, vegetables, whole grains, lean protein, and healthy fats is fundamental (4). Reducing sodium, sugar, and saturated fat intake supports better cardiovascular health and blood pressure control (4).
- **Stress and Mental Health Management:** Managing stress through relaxation techniques like meditation and yoga can help minimize its impact on cardiac health (4). Adequate sleep and access to counselling or therapy are encouraged for emotional well-being (4).
- **Recognizing Symptoms:** Educating patients to identify and report symptoms such as chest discomfort, fainting, or worsening breathlessness ensures timely medical intervention, preventing complications (4).



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A Rare Case of Anterior Staphyloma with a Painful Blind Left Eye treated successfully at Aster Hospital, Al Qusais

Dr. Faizan Mehmood
Ophthalmology (Specialist)

PRESENTATION

- 42-year-old female
- Surgical history of kidney surgery 2 years back, followed by sudden decreased vision in the left eye
- Family history of Diabetes
- Admitted with Painful Blind Eye - no perception of light (PL)

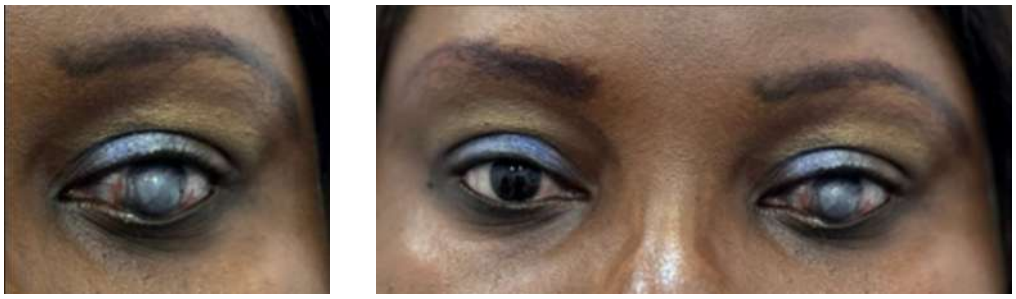
FINDINGS

During Examination:

- Visual Acuity with glasses: Right eye - 6/6; Near vision - N6; Left eye – PL negative
- Retinoscopy in right eye: clear glow
- PGP (progressive): Right eye +0.00ds/-0.50dcx50; NV add +1.50ds
- IOP - Right eye: 12 mmHg
- Slit lamp evaluation: RE WNL; Cornea Clear; PNSNR; Central fundus WNL

Left Eye Examination:

- Anterior staphyloma
- Conjunctival congestion
- Total corneal macular opacity
- PL negative
- Ocular Motility intact



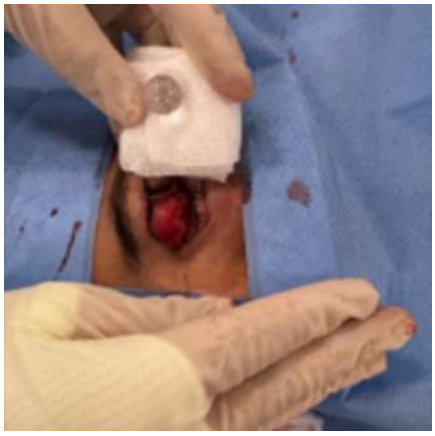
Pre-op images showing the affected Left Eye

DURING PROCEDURE

The patient underwent Left Eye Evisceration with a Ball Implant followed by fitting of Ocular Prosthesis:

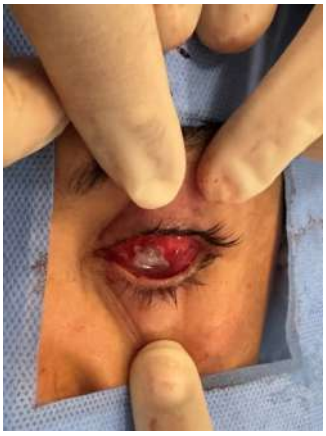
- Local anaesthesia was given, and parts were cleaned and draped.
- 360-degree conjunctival peritomy was done.
- After sub-conjunctival dissection, a stab incision at the limbus was given with 11 no. blade.
- Cornea was excised with corneal scissors.
- Intraocular contents were scooped with an evisceration scoop.
- Haemostasis was achieved with cotton packing.
- Polymethylmethacrylate (PMMA) ball of 20mm size was inserted, and the sclera was closed with 6-0 Vicryl.
- Conjunctiva was closed with 6-0 Vicryl with continuous sutures.
- And a conformer was placed.

Intra-op Images



Eviscerated Eye with PMMA Ball

Scleral and Conjunctival Closure with Ball inside, conformer to be placed inside the eye



Final pic of the procedure showing the ball and conformer in place

POST PROCEDURE

The patient tolerated the procedure well and was in a stable condition on discharge. Regular follow-up was done. A customized ocular prosthesis was made and fitted after 1 month of surgery with excellent cosmesis.

Post-op Image

DISCUSSION

Introduction:

Evisceration of the eye is a surgical procedure involving the removal of intraocular contents while preserving the sclera and extraocular muscles. It is a less invasive alternative to enucleation, offering better cosmetic outcomes and quicker recovery. The primary aim of this procedure is to alleviate pain and improve the patient's quality of life in cases where the eye has lost functional and structural integrity.

Indications:

Evisceration is indicated in conditions such as:

- Severe trauma leading to irreparable damage
- Endophthalmitis unresponsive to treatment
- Painful blind eyes due to glaucoma or phthisis bulbi
- Severe cosmetic disfigurement

It is contraindicated in suspected intraocular malignancy or cases with a high risk of orbital spread of infection.

Surgical Technique and Implants:

Modern evisceration techniques focus on preserving orbital volume and enhancing prosthetic mobility. Orbital implants made of materials like hydroxyapatite and porous polyethylene have significantly reduced complications like implant migration and exposure. A customized prosthesis ensures better cosmetic results and patient satisfaction.

Advantages:

- Preserved scleral shell allows for better prosthesis motility and appearance
- Lower psychological impact due to preserved anatomical structures

Challenges:

- Risk of implant extrusion or infection
- Inappropriate for patients with intraocular malignancies

Outcomes:

Studies consistently report high levels of cosmetic satisfaction and pain relief post-evisceration:

- 92% success rate in achieving satisfactory prosthesis fit
- A significant improvement in patients' quality of life after orbital implantation

CONCLUSION

Evisceration is a valuable surgical option for blind, painful eyes, balancing effective pain relief with improved cosmetic outcomes. Advances in surgical techniques and implant technologies continue to enhance the safety and efficacy of this procedure.

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