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Heart-to-Heart: Cardiovascular Palliative Care in An Adolescent with Rheumatic Heart Disease & Thoracoabdominal Aortic Aneurysm

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Background

• Rheumatic heart disease (RHD), results from valve damage from acute rheumatic fever, primarily affects the mitral valve, leading to stenosis or regurgitation. As the disease progresses, it causes heart failure symptoms and often necessitates valve surgery if with severe valvular dysfunction, especially if the patient is symptomatic. While the optimal timing for surgery remains uncertain, recent evidence supports early intervention—before irreversible myocardial damage or severe valve changes—particularly in young patients with a longer life expectancy, to improve surgical outcomes. In the Elegant study of patients aged 5 to 24 years with RHD, 16.2% were diagnosed with severe disease; and among these, 50% had undergone valve surgery within two years, and 10% had died within six years.[1]

• Thoracoabdominal aortic aneurysm (TAAA) results from progressive dilation of the descending thoracic aorta extending into the abdominal aorta, with potential complications such as rupture or dissection. Symptoms may include chest, back, or abdominal pain, shortness of breath, or hypotension.[2] Early studies recommend repair in symptomatic patients and before the aneurysm reaches 7.0 cm. Open surgical repair remains the definitive treatment due to its long-term durability, particularly in patients with a life expectancy over 10 years, but carries significant risks, including death, spinal cord injury, renal failure requiring dialysis, and stroke.[3]

• Both surgical management – open TAAA repair and mitral valve come with both benefits and risks.

Purpose

• The case highlights cardiovascular palliative care in a patient with TAAA and RHD w/ severe mitral valve regurgitation who preferred conservative management over surgical repair for both disease.

Case Description

• An 18/F, single, college student, admitted under the Pediatric service due to fever and desaturations, patient was diagnosed with PCAP, non-severe 3 weeks prior.

• Past medical history: RHD (severe MR, mild AI, severe TR) diagnosed 7 years ago, on Enalapril, Dioxin, Furosemide, Benzathine penicillin, with regular follow-up

• Born full term to a then 24-year-old G3P2 (2002) mother via SVD, with no fetomaternal complications

• Independent to all basic and instrumental ADLs prior to admission

• PE: underweight, tachypnea, occasional bilateral rhonchi, dynamic precordium, grade 4/6 holosystolic murmur, heaves, non-tender abdomen, hepatomegaly

Imaging

• **Vascular POCUS**: suprarenal fusiform abdominal aortic aneurysm with thrombus formation, normal common carotid, internal carotid, and external carotid arteries

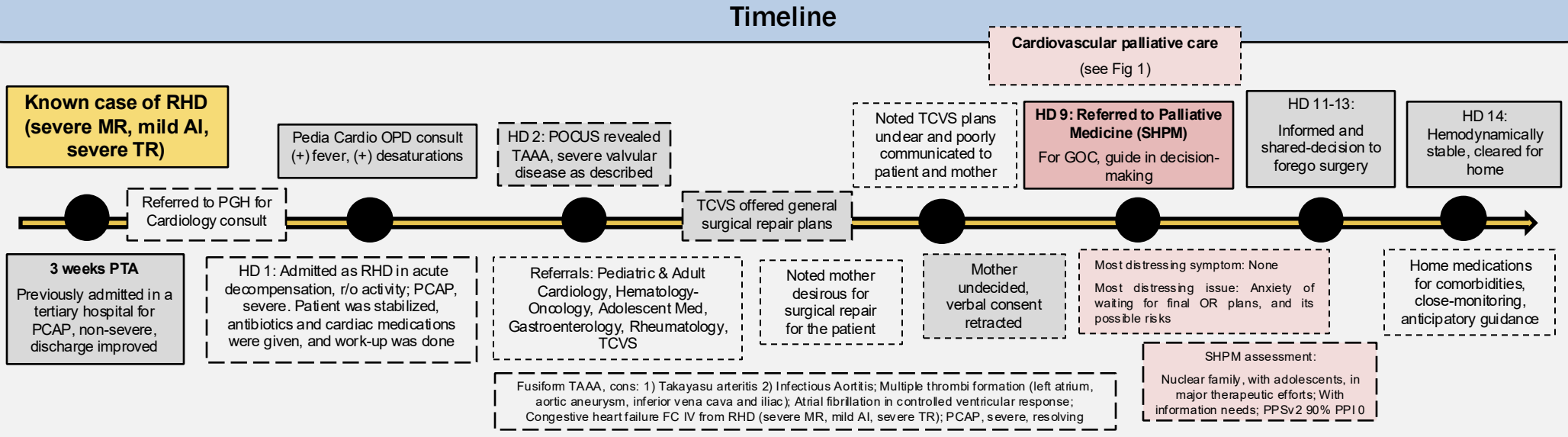
• **2D Echo:** Mitral valve: *thickened leaflets, fixed PMVL with limited mobility, prolapsing AMVL, leaflets poorly coaptating causing severe mitral regurgitation;* Tricuspid valve: *leaflets appear normal and with good mobility, moderate to severe tricuspid regurgitation;* Aortic valve; *minimal thickening of aortic valve leaflets with central gap in coaptation causing mild aortic regurgitation; Left atrial thrombus measuring 5.4cm x 5.4 cm; Left atrial enlargement, left ventricular enlargement; Preserved biventricular systolic function; Consider mural thrombus on abdominal aorta; Atrial fibrillation in CVR during study*

• **CT Angiography:** *Thoraco-abdominal fusiform aortic aneurysm with thrombus formation. No evidence of aortic dissection or rupture (5.1 cm x 4.8 cm W x AP)*

Multi-chambered cardiomegaly. Consider interior vena cava and iliac thrombosis.

Apical findings and granulomata formation are suggestive of a chronic inflammatory process. Subsegmental and segmental atelectasis vs non-specific parenchymal fibrosis, bilateral

Timeline



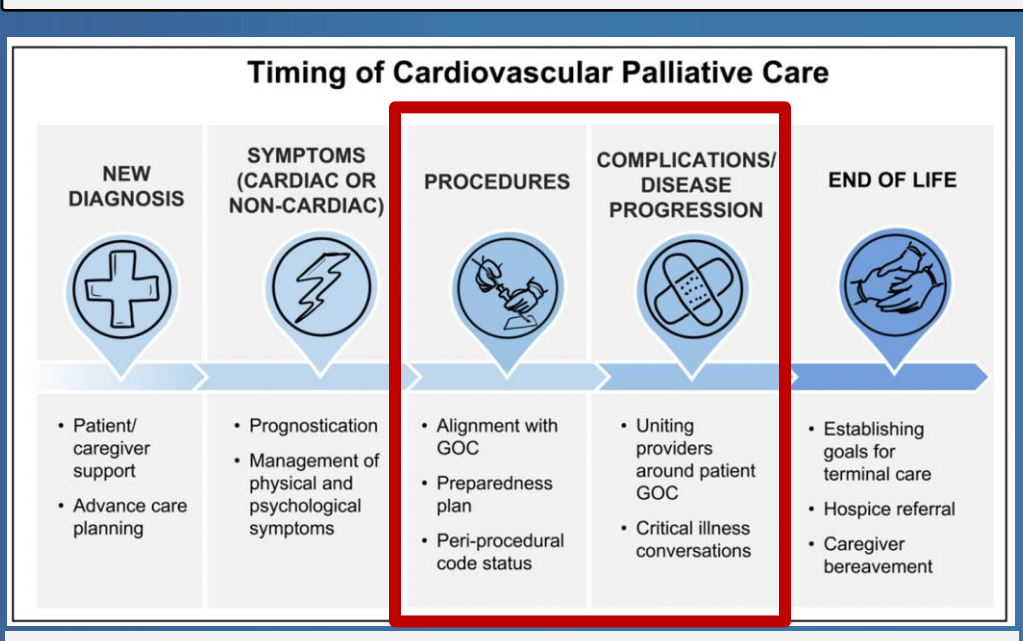


*Fig 1. Timing of palliative care throughout cardiovascular disease course. GOC indicates goals of care (*from Godfrey et al., 2023)

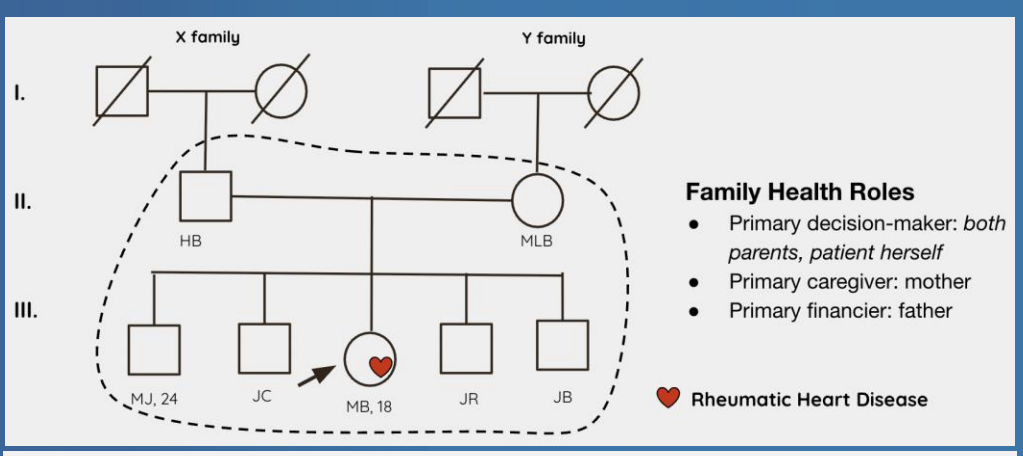


Fig 2. Family genogram and family health roles

Palliative Management

• Effective communication was employed during the dialogues with the mother and the patient. Allowing them to express feelings and verbalize concerns.

• Family health roles were identified (fig 2).

• The most distressing symptom and issue were identified, as well as the goals of care.

• Family was allowed to make an informed decision after coordination with TCVS to discuss clearly the planned surgeries, in terms of procedures, risks and benefits.

• An informed and shared decision involving the patient and family and the whole care team congruent to the goals of care was ensured.

• Referral to Nutrition service for upbuilding was done.

• Psychoemotional support rendered.

• Anticipatory care planning initiated with mother. Advance care directives introduced.

Discussion

• Cardiovascular palliative care is a comprehensive, team-based approach designed to enhance quality of life throughout the entire course of cardiovascular disease—from diagnosis to end of life—by focusing on symptom relief, effective communication to determine care goals, and offering psychosocial and spiritual support (fig 1).[5]

• Palliative care for individuals with valvular heart disease often emphasizes advance care planning and collaborative decision-making regarding the appropriateness and timing of valvular interventions.[5]

• Cardiovascular palliative care for aortic aneurysms aims to enhance the quality of life for patients with advanced disease, particularly those who are not candidates for surgical intervention or opt against surgical intervention.[6]

Insights

•Cardiovascular palliative care may be integrated at any stage in the natural progression of cardiovascular disease.

• Incorporating palliative care for patients who decline repair for abdominal aortic aneurysm and severe valvular heart disease facilitates alignment of treatment with their goals throughout the course of care.

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