



Anesthetic Management of Severe HOCM with Fixed RV Compression from Pectus Excavatum in a Prone Cervical Spine Surgery

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INTRODUCTION

Costello Syndrome is a rare *RASopathy* that presents with craniofacial features, developmental delay, failure to thrive and **cardiac abnormalities**, that usually include arrhythmias and Hypertrophic Cardiomyopathy (HOCM)¹.

CLINICAL CASE

27 year-old male with Costello syndrome who presented with left-sided weakness and gait/balance problems in the setting of cervical stenosis and myelomalacia

HOCM s/p LVOT resection (LVOT gradient 90mmHg)

Severe MR

Pectus Excavatum with RV compression

Chiari malformation with VP shunt

Multiple prior spine surgeries

Urgent C2- C4 laminectomy and C2-C6 fusion

HEMODYNAMICS CONSIDERATIONS

HCOM – goals to prevent LVOT obstruction ²

Maintain SVR

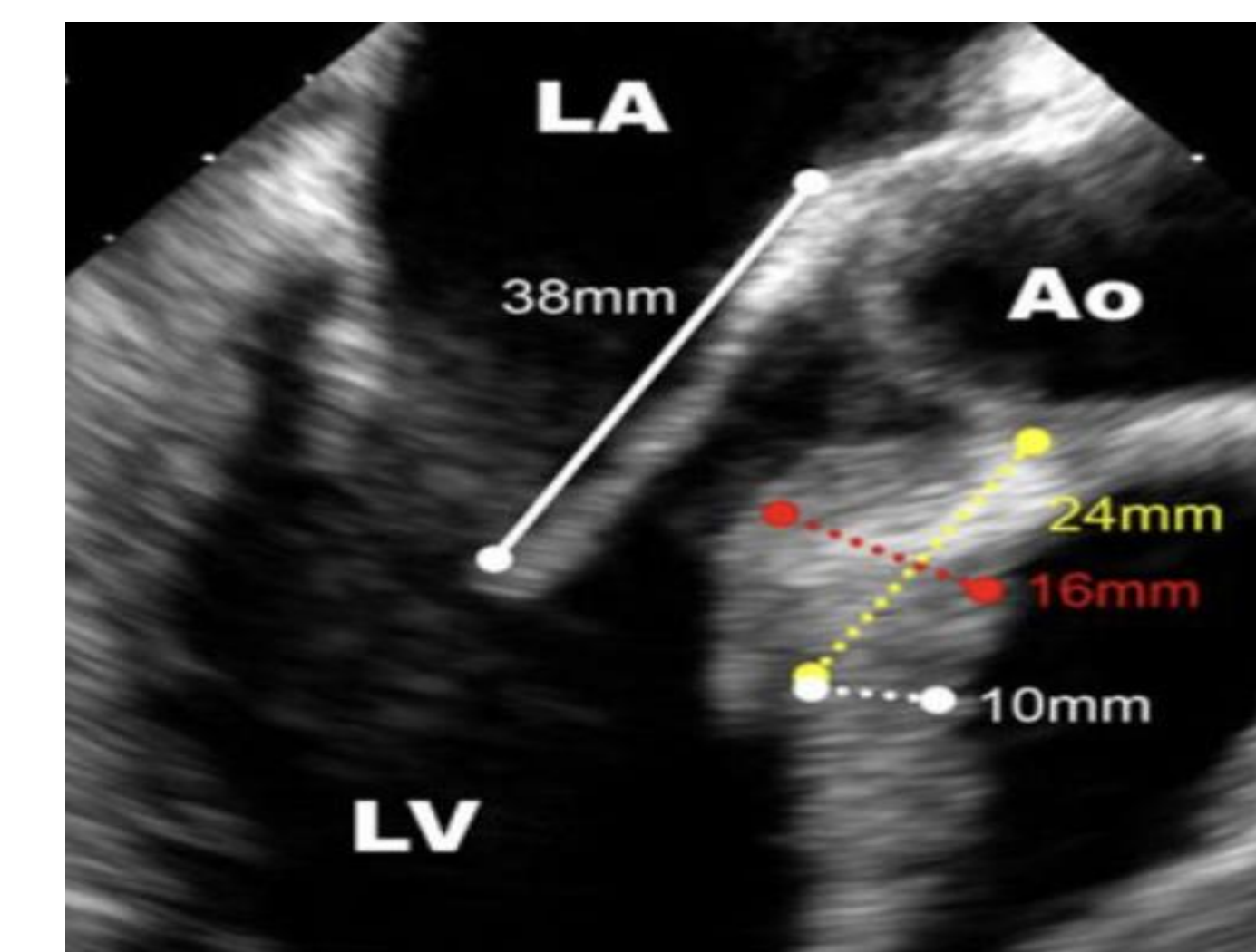
Maintain Preload

↓HR – allow LV filling

Challenges

- Pectus excavatum poses **competing goals**: RV filling limited by chest wall compression → **↑HR to maintain LV filling.** ^{3, 4}
- **Preload** is significantly reduced in the setting of fixed RV compression.
- Prone positioning can **worsen RV compression.** ⁵
- The use of TTE for intraoperative management guidance is **technically difficult in prone** positioning ⁶, especially in neck surgery.

IMAGES



CONCLUSIONS

Managing hemodynamics in patients with **competing physiologic goals** for modifiable factors (HR, SVR, fluid resuscitation) can be extremely challenging.

TEE played a key role in intraoperative decision-making by providing real-time assessment of right ventricular (RV) compression after prone positioning.

TTE is **rarely performed prone position** due to technical challenges.

Team collaboration between anesthesia and surgical teams was crucial for adapting intraoperative plans.

Pre-established **backup plans** are essential for safely managing high-risk cases with challenging hemodynamics.

ANESTHETIC MANAGEMENT

Adequate access
1L pre-operatively
Arterial line

Induction

First TEE
assessment prior
to prone

Hemodynamic **trial of prone** positioning under
TEE guidance
RV compression remained stable
Decision to proceed with the surgery

- ✓ MAP > 85mmHg with use of vasopressors
- ✓ HR < 80bpm with beta-blockade
- ✓ Limit surgical time with orthopedic surgery
- ✓ IVF failed to improve RV preload on TTE – limited IVF to 3L total

Tolerated
prone and
anesthetic

Extubated to
NC 6L

Transported
to CT ICU

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