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ANESTHETIC EXPERIENCE OF INFANT WITH LV NONCOMPACTION (SPONGIFORM CARDIOMYOPATHY) - A CASE REPORT

S. Aswar, Alder Hey Children's Hospital, UK

INTRODUCTION

Left ventricular noncompaction (LVNC) is a rare, congenital cardiomyopathy with a poor long-term prognosis. The main manifestations are global left ventricular (LV) dysfunction, arrhythmias, systemic emboli, and eventual heart failure. Therefore a special precaution is warranted before administrating general anesthesia.

CASE REPORT

A 4 month-old boy diagnosed to have moderate LV dysfunction: dilated thickened non-compacted LV myocardium with EF 30% along with mild MR, and since then he was on anti-failure medication. For grade III gastro-esophageal reflux he was posted for laparoscopic gastrostomy with fundoplication. Injection dobutamine infusion was started (3ug/kg/min) in view of hemodynamic instability

We planned GA. Continuous monitoring of blood pressure was initiated after a right femoral arterial catheter. Anesthesia was induced with fentanyl 4 mcg/kg and rocuronium 1.2mg/kg (modified rapid sequence induction), followed by intubation. Anesthesia was maintained with inhalation. Although induction was uneventful, BP abruptly dropped from 76/49mmHg to 55/39mmHg which was treated with boluses of injection phenylephrine. The patient was stable for the rest of the procedure. Awakening and recovery were smooth after the procedure followed by on-table extubation.

DISCUSSION

Diagnosis of LVNC is based on the echocardiographic findings: prominent trabeculation, deep intertrabecular recesses, and blood flow into the intertrabecular recesses.

The clinical presentation of patients with LVNC is variable including congestive heart failure, arrhythmias, and systemic thromboembolic events. The current recommendations for management include beta-blockers, ACE inhibitors, and diuretics.

The anesthetic management of these patients requires specific attention to heart failure, arrhythmia, thromboembolic events, and NMDs. TEE, if available, is a useful monitor to guide intraoperative anesthetic and fluid management in the presence of severely depressed LV and RV function. Beat to beat intra-arterial blood pressure monitoring is highly recommended.

CONCLUSION

The anesthesiologist should be specifically concerned with the presenting symptoms of LVNC, heart failure, thromboembolic events, arrhythmia and NMDs.

References:

1. Chen R, Tsuji T, Ichida F. Mutation analysis of the G4.5 gene in patients with isolated left ventricular noncompaction. Mol Genet Metab. 2002;77:319–325. [PubMed]

2. Ichida F, Tsubata S, Bowles KR. Novel gene mutations in patients with left ventricular noncompaction or Barth syndrome. Circulation. 2001;103:1256–1263. [PubMed]

3. Oechslin EN, Attenhofer Jost CH, Rojas JR. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. J Am Coll Cardiol. 2000;36:493–500. [PubMed]