

Complete Cleft Sternum and Tetralogy of Fallot in a Newborn

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Introduction: Complete cleft sternum (CCS) is a rare disease wherein sternal agenesis leads to total nonunion of the anterior ribs at the midline. CCS is often discussed in association with other sternal defects such as partial sternal cleft and ectopia cordis, although it is a distinct disease entity. In contrast with certain forms of ectopia cordis, isolated CCS usually has a favorable outcome when repaired early in life¹. Until recently, CCS was not associated with cardiac anomalies. However, 3 case reports now exist of CCS with a cardiac defect: 1) double outlet right ventricle and ventricular septal defect²; 2) patent ductus arteriosus³; and 3) pulmonary valve stenosis with conoventricular and atrial septal defects⁴. A fourth case describes a patient with a partial (superior) sternal cleft and tetralogy of Fallot⁵ (TOF). We herein report the anesthetic management of an infant with CCS and TOF.

Case Report: A 2.5 kg term female was diagnosed perinatally with CCS, TOF, and a single left-sided superior vena cava. On day 3 of life, the baby was intubated secondary to respiratory distress thought to be due to flail chest. She was brought to the operating room on day 4 of life for cleft sternum repair. The baby's heart was grossly discernable as it contracted beneath the skin. The skin demonstrated a supraumbilical midline raphe (see photo). Anesthesia was induced with fentanyl and maintained with isoflurane and pancuronium. A posterior tibial arterial line was placed after induction. The surgeons then proceeded to expose the heart. The subcutaneous tissues were undermined extensively. During this time, the infant was ventilated with 22% oxygen and peak inspiratory pressures of 21 mm Hg. Systolic blood pressures averaged 70 – 78 mm Hg, oxygen saturation ranged from 85 – 93%, and arterial blood gas values were pH=7.43, PCO₂=42, PO₂=62. When free rib margins were approximated in the midline with suture forming a neosternum, systolic blood pressures fell to a nadir of 55 mm Hg, the inspired oxygen was increased to 35% to maintain stable oxygen saturation, and peak inspiratory pressures were increased to 26. At this time, blood gas values were pH=7.34, PCO₂=53, PO₂=61. Systolic blood pressure was maintained by bolus administration of 5% albumin. Total fluids for the case were 5% albumin (24 ml/kg) and Lactated Ringer's solution (24 ml/kg). The patient was transferred intubated, ventilated, and sedated to the cardiac intensive care unit uneventfully. On post-operative day 10 the patient returned to the operating room for revision of sternal closure with sternal wires following partial wound dehiscence. She is currently awaiting definitive repair of her TOF.

Discussion: This case combines the rare finding of a CCS with TOF, a previously unreported congenital association. Repair of simple CCS requires special attention to the potential for mechanical cardiac compression during approximation of the overlying bony structures, but nevertheless is associated with very good outcomes. The potential for hemodynamic instability is compounded by coexisting congenital heart disease. In this patient with TOF, there was only mild right ventricular outflow tract obstruction, and she was thus able to maintain adequate pulmonary blood flow. Nevertheless, her cardiac output was considerably stressed by the surgical repair, but not to the point of requiring vasoactive support.

Conclusions: Anesthesia was safely provided for the challenging closure of a CCS in a patient with TOF. Stability was achieved through careful attention to invasive hemodynamic monitoring and maintenance of cardiac preload. A margin of safety was enjoyed due to the absence of any significant right ventricular outflow tract obstruction.



Figure 1: Cleft sternum, pre-operative

References

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