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A 7-year-old male with Prader-Willi Syndrome and cyanotic heart disease presented with fatigue and abdominal pain. His cardiac anatomy consisted of tricuspid atresia, d-transposition of great arteries, ventricular septal defect, and a straddling mitral valve with Fontan circulation. His comorbidities included atrioventricular block with an epicardial pacemaker, cerebrovascular accident, and obesity (Figure 1). A diagnosis of acute cholecystitis was obtained by ultrasound demonstrating pericholecystic fluid, a thickened gall bladder wall, and cholelithiasis (Figure 2).

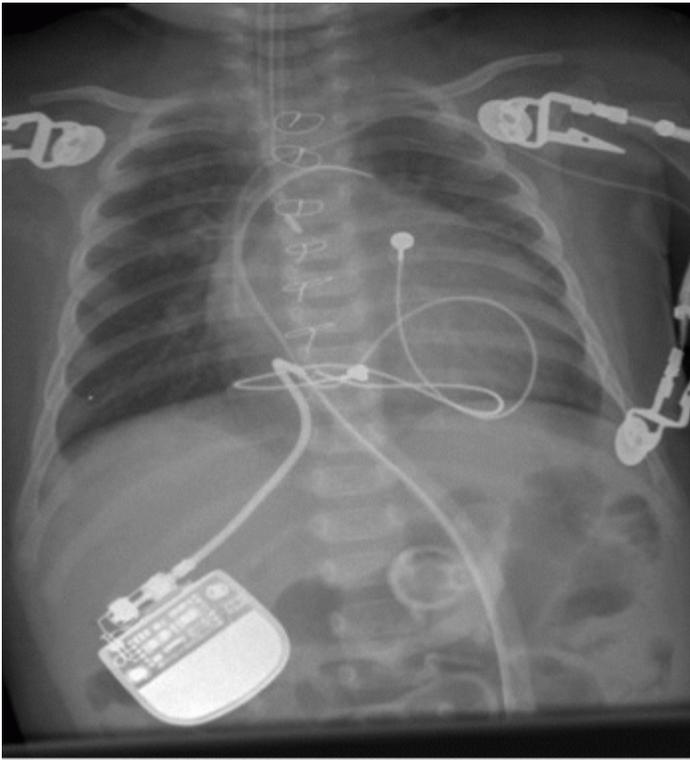
Prior to the OR, his pacemaker was set in asynchronous mode. Anesthesia was induced with fentanyl 2 mcg/kg, ketamine 2 mg/kg, and vecuronium 0.1 mg/kg with preserved hemodynamic stability. Easy bag mask ventilation was confirmed, and laryngoscopy revealed a grade I view. Standard monitors, an arterial line and central line were placed. He tolerated abdominal insufflation and surgery well (Table 1).

Prader-Willi Syndrome is due to partial deletion of chromosome 15. Infants with Prader-Willi exhibit developmental delay, and poor feeding (1,2). After age 5, hyperphagia, obesity, sleep apnea, and pulmonary hypertension can occur. Associated airway concerns include: high arched palate, micrognathia and decreased functional residual capacity (3).

Congenital cardiac diseases associated with Prader-Willi are commonly limited to benign arrhythmias. In our patient, however, single ventricle physiology coexisted. Creation of a carbon dioxide pneumoperitoneum impacts Fontan physiology by increasing intra-abdominal pressure and pulmonary vascular resistance which ultimately decreases cardiac output. Despite these risks multiple Fontan patients have tolerated laparoscopy (4, 5).

Patients with Prader-Willi Syndrome and Fontan physiology represent various challenges to the anesthesiologist. Minimizing respiratory and cardiovascular compromise requires optimizing preoperative preparation, limiting airway manipulation, and effects of laparoscopy.

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Time	Stage of operation	FiO2	End-tidal CO2	Peak Inspiratory Pressure	Central Venous pressure	Respiratory Rate	HR	SBP	DBP	MAP	Arterial blood gas
1742	Pre-incision	74%	38	17-19	19 cmH2O	9-12	90	100-125	55-80	68-90	7.33/45/189
1841	Pre-insufflation	72%	39	17-19	20 cmH2O	12-14	90	111-122	64-68	78-85	7.27/51/235
1856	Insufflation	74%	42	18-20	21 cmH2O	16-18	90	118-127	65-78	81-92	
1941	Post-Insufflation	80%	35	19-23	≤ 20 cmH2O	16-18	90	126-135	67-80	80-98	7.29/46/294