A 7 year old male with a history of Prader-Willi Syndrome and complex cyanotic congenital heart disease presented to our tertiary care center with symptoms of fatigue and right upper quadrant abdominal pain exacerbated by defecation. His cardiac anatomy consisted of: tricuspid atresia, d-transposition of great arteries, ventricular septal defect, and a straddling mitral valve. He was maintained on aspirin, for which he received an epicardial pacemaker, cerebrovascular accident with residual left sided weakness, subclinical hypothyroidism, morbid obesity, undescended testes, growth failure and type two diabetes mellitus. He was extubated post-operatively in the operating room.

Figure 2. Transverse ultrasonic view of gallbladder shows thickened wall and calculi.

Induction proceeded with fentanyl 2 micrograms per kilogram, ketamine 2 mg/kg, vecuronium 0.1 mg/kg and isoflurane 1% with preserved hemodynamic stability. Easy bag mask ventilation was confirmed, and a 5.5 endotracheal tube was placed with a Miller 1.5 laryngoscope and a grade I view was noted. In addition to standard ASA monitors, an ultrasound-guided 20 gauge right arterial line and a 4 French right internal jugular central venous line was placed. He had a stable intraoperative course, tolerating abdominal insufflation, (Figure 3) and was extubated post-operatively in the operating room.

<table>
<thead>
<tr>
<th>Time</th>
<th>Stage of operation</th>
<th>FO2</th>
<th>End tidal CO2</th>
<th>Peak Inspiratory Pressure</th>
<th>Central Venous pressure</th>
<th>Respiratory Rate</th>
<th>HR</th>
<th>SBP</th>
<th>DBP</th>
<th>MAP</th>
<th>Arterial blood gas</th>
</tr>
</thead>
<tbody>
<tr>
<td>1742</td>
<td>Pre-    infusion</td>
<td>74%</td>
<td>38</td>
<td>17-19</td>
<td>19 cmH2O</td>
<td>9-12</td>
<td>96</td>
<td>100</td>
<td>125</td>
<td>60</td>
<td>66-80</td>
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<tr>
<td>1841</td>
<td>Post-   infusion</td>
<td>74%</td>
<td>39</td>
<td>17-19</td>
<td>19 cmH2O</td>
<td>12-14</td>
<td>96</td>
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<tr>
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<td>42</td>
<td>16-20</td>
<td>21 cmH2O</td>
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<tr>
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<td>16-18</td>
<td>96</td>
<td>95</td>
<td>80</td>
<td>65</td>
<td>60-88</td>
</tr>
</tbody>
</table>

Figure 3 represents hemodynamic and respiratory data during different phases of our patient’s intraoperative course. HR = heart rate, SBP = systolic blood pressure, DBP = diastolic blood pressure, MAP = mean arterial pressure.

Discussion

Prader-Willi Syndrome is due to partial deletion of chromosome 15. As neonates, these patients exhibit hypotonia due to fatty infiltration of fast twitch muscle fibers (MacKenzie), developmental delay, poor feeding and failure to thrive. In contrast, after age 5 they are overweight due to hypothalamic-mediated hyperphagia (Zipf). Eventually, obesity and obstructive sleep apnea lead to chronic hypoventilation and pulmonary hypertension. Airway concerns for these patients include: high arched palate, micrognathia and decreased functional residual capacity (Gregory). These concerns are exemplified by one case of post-operative sleep apnea that persisted for days and was associated with bradycardia and an oxygen saturation nadir of 85% (Dearlove).

Congenital cardiac diseases associated with Prader-Willi are commonly limited to arrhythmias such as premature ventricular contractions. In this patient, however, tricuspid atresia with d-transposition and ventricular septal defect coexisted. This was managed ultimately with Fontan circulation. Fontan physiology is the final palliative stage for many single ventricle lesions. Pulmonary blood flow is passive and is easily altered by pulmonary vascular resistance and preload, specifically the transpulmonary gradient, which is the difference between central venous pressure and systemic ventricular end-diastolic pressure (Naeije).

Pneumoperitoneum created during laparoscopic surgeries has many hemodynamic consequences for patients with Fontan physiology. Carbon dioxide insufflation increases intra-abdominal pressure which increases intrathoracic pressure, ultimately raising mean airway pressure and impeding systemic venous return and pulmonary blood flow. Decreased pulmonary blood flow decreases cardiac output. In the case of a fenestrated Fontan, cardiac output may be maintained at the expense of oxygen saturation. Additionally, absorbed carbon dioxide can increase pulmonary vascular resistance and again decrease pulmonary blood flow. Despite these theoretical risks multiple case reports of patients with Fontan physiology have undergone laparoscopy safely (Taylor, McClain).

References