Perioperative Management of an Infant with Congenital Marfan’s Syndrome

Justinn Tanem, M.D. and John P. Scott, M.D.
Department of Anesthesiology, Medical College of Wisconsin
Milwaukee, WI

CASE PRESENTATION

We present the case of a 41 day-old, ex-36 7/12 GWA male infant requiring exploratory laparotomy for persistent feeding intolerance and emesis. The infant was born weighing 2515 grams via Cesarean section to a 31 year-old, G3P0 mother. The prenatal course was complicated by fetal SVT, managed with digoxin and flecainide. He subsequently developed biventricular failure and AV valve insufficiency due to persistent atrial flutter prompting the decision to deliver at 36 weeks to further manage his tachycardia.

Following delivery he was intubated and noted to have wide-complex tachycardia necessitating defibrillation and administration of amiodarone, with successful conversion to sinus rhythm. The child continued to have intermittent tachycardia prompting the initiation of digoxin. He had persistent respiratory insufficiency secondary to his pectus excavatum and necessitated high-flow nasal cannula with increasing FiO2 requirements.

Due to feeding intolerance, the infant was taken to the operating room on day of life 13 for a Ladd’s procedure with lysis of adhesions. Post-operatively, a diagnosis of congenital Marfan’s syndrome was confirmed by exome sequencing, which demonstrated a novel FBN1 variant. Due to persistent feeding intolerance the child was maintained on TPN and subsequently presented to the operating room for exploratory laparotomy, gastrostomy tube placement, and open pyloroplasty on day of life 41.

Intraoperative

Standard ASA and NIRS monitors were utilized. Patient underwent intravenous induction with propofol, fentanyl, and rocuronium. The infant was intubated via direct laryngoscopy, with a grade I view. A single-shot caudal epidural was administered with bupivacaine and morphine for analgesia and to mitigate sympathetic stimulation, tachycardia, and aortic stress in the setting of significant aortic dilation. The procedure was completed with minimal blood loss. The patient received 50 cc/kg of crystalloid in addition to TPN. The anesthesiologist noted aortic root dilation and systolic hypertension, which may increase regurgitant fraction.

Fig. 1: Photograph of infant demonstrating pectus deformity and overlapping digits

Fig. 2: Echocardiogram demonstrating severely dilated aortic root

Marfan’s syndrome is a connective tissue disorder caused by mutations of the FBN1 gene on chromosome 15 encoding fibrillin-1, a key component of the extracellular matrix. It is inherited in an autosomal dominant manner with an incidence of approximately 1 in 5000 individuals with complete penetrance and variable expression. Clinical diagnosis is made via the Ghent Criteria.

Table 1: Ghent criteria, 2010

<table>
<thead>
<tr>
<th>Absent family history</th>
<th>Aortic root Z score of &lt;2 and ectopia lentis</th>
<th>Aortic root Z score of ≥2 and an FBN1 mutation</th>
<th>Aortic root Z score of ≥2 and a systemic Z score of ≥2</th>
<th>Ectopia lentis AND an FBN1 mutation with a known aortic pathological abnormality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive family history</td>
<td>Ectopia lentis</td>
<td>Systemic Z score of ≥2</td>
<td>Aortic root Z score of ≥2</td>
<td>Absent family history AND multiple organ system involvement</td>
</tr>
</tbody>
</table>

Table 2: Anesthetic considerations in congenital Marfan’s Syndrome

Congenital Marfan’s syndrome is a rare disorder that affects multiple organ systems and poses numerous challenges to the anesthesiologist. Care must be taken to maintain appropriate hemodynamics, prepare for possible difficult intubation, and mitigate joint dislocations and soft tissue injuries.

REFERENCES

Table 1: Ghent criteria, 2010

Clinical abnormality | Anesthetic consideration
<table>
<thead>
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<tbody>
<tr>
<td>Aortic root dilation</td>
<td>Avoid tachycardia and hypertension, which may increase regurgitant fraction</td>
</tr>
<tr>
<td>Polyvalvular insufficiency</td>
<td>Avoid bradycardia and hypotension, which may increase regurgitant fraction</td>
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<tr>
<td>Airway:</td>
<td>May present with difficult intubation</td>
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<tr>
<td>Respiratory:</td>
<td>May present with difficult intubation</td>
</tr>
<tr>
<td>Musculoskeletal:</td>
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