Anesthetic Management of a Neonate with Jeune Sydnome

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BACKGROUND

Jeune syndrome (asphyxiating thoracic dystrophy) is a rare autosomal recessive chondrodysplasia that presents multiple challenges to the anesthesiologist. Although this is predominantly due to abnormalities including thoracic and lung hypoplasia, there are other factors to consider when anesthetizing patients with this disorder. We present a 7- day old infant with Jeune syndrome that underwent MRI imaging to evaluate a cerebral cyst.

CASE REPORT

Past Medical History

- Twin B of a di/di twin gestation born at 36 weeks via C-section due to pre-eclampsia and breech presentation
    - IUGR, birth weight: 1870 grams
- Bradycardia (HR 80) and respiratory distress at birth
- Apgar scores: 3, 6, 7 at 1, 5 and 10 minutes
- Increasing FiO2 requirements and severe acidosis with a pH of 6.79 on CPAP
- Endotracheal intubation shortly after birth
- Imaging demonstrated a narrow thoracic cage with small lung volumes (fig 1), various skeletal deformities (fig 2) and renal dysplasia
- TTE: small patent foramen ovale with a left-to-right shunt, a small congenital VSD, increased right ventricular size and wall thickness with increased right systolic pressures, reduced right systolic function and pulmonary hypertension.

IMAGES

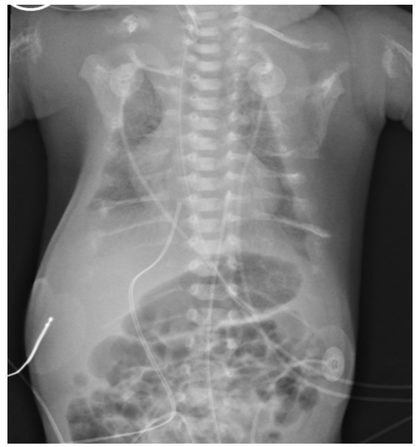

Figure 1. Chest x-ray after birth demonstrated a small, narrow thorax with low lung volumes, bilateral streaky airspace opacities and shortening of the ribs.

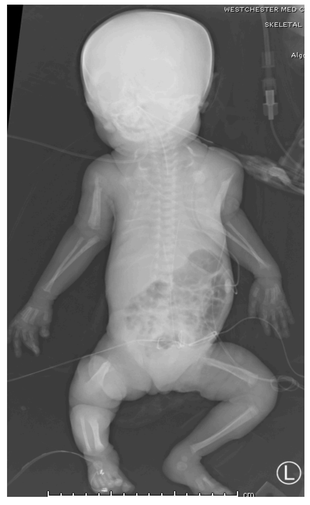

Figure 2. Skeletel survey demonstrated rhizomelic shortening, as well as deformities and decreased ossification of the distal ribs, distal clavicles and proximal femurs and humeri.

CASE REPORT (Cont.)

Anesthetic Course

- Presented for Brain MRI for evaluation of cerebral cyst
- Remained on mechanical ventilation via a 3.0 uncuffed endotracheal tube
- Induced and maintained on Sevoflurane at 0.8 MAC; no other IV medications administered
- Pressure-controlled mechanical ventilation utilized to deliver low tidal volumes
- Hemodynamically stable on an FiO2 of 50%
- MRI completed without any complication; patient remained intubated

CONCLUSIONS

Jeune syndrome is a rare autosomal recessive chondrodysplasia that presents multiple challenges to the anesthesiologist:

- Respiratory insufficiency and recurrent pulmonary infections can result in respiratory failure and death in infancy
- Patients have small lung volumes, decreased FRC and decreased airway compliance
- High airway pressures should be avoided and low tidal volumes with increased inspiratory times are preferred to prevent barotrauma and pneumothorax
- Skeletal deformities require careful intraoperative positioning to avoid nerve injury
- Systemic manifestations include hepatic, pancreatic, bile duct and renal anomalies and low doses of anesthetic medications should be utilized secondary to impaired hepatic and renal function
- Lung hypoplasia can result in pulmonary hypertension with subsequent heart failure

REFERENCES

Mehmet I. Buget, Emine Ozkan, Ipek S. Edipoglu, and Suleyman Kucukay, “Anesthetic Approach for a Patient with Jeune Syndrome,” Case Reports in Anesthesiology, vol. 2015, Article ID 509196, 3 pages, 2015. doi:10.1155/2015/509196

Saletti D, Grigio TR, Tonelli D, Ribeiro OD, Marini F. Case report: anesthesia in patients with asphyxiating thoracic dystrophy: Jeune syndrome. RevBrasAnestesiol. 2012;62(3):424-431. http://dx.d0i.0rg/l 0.1016/S0034-7094(12)70142-3.

L. M. Borland, “Anesthesia for children with Jeune's syndrome (asphyxiating thoracic dystrophy),” Anesthesiology, vol. 66, no. 1, pp. 86–88, 1987