

A Case Report of a Cesarean Section Parturient with Cantu Syndrome

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Introduction

Cantu syndrome is a rare autosomal dominant genetic disorder characterized by hypertrichosis, coarse facial features, cardiac abnormalities, and skeletal abnormalities and can be associated with mutation of the ABCC9 gene.

Common Features of Cantu Syndrome

- Facial anomalies
 - low-set ears
 - exophthalmos
 - broad flat nasal bridge
 - large mouth and thick lips
 - macroglossia
- Cardiovascular anomalies
 - cardiac enlargement
 - patent ductus arteriosus
 - aortic root dilation
 - pericardial effusion
- Hypertrichosis
- Skeletal abnormalities
 - frontal bossing
 - osteochondrodysplasia including short neck
 - thickened calvarium
 - wide vertebral bodies
 - wide ribs
 - hypoplastic pelvis



Figure 1. CXR demonstrates rods from spinal fusion and lumboperitoneal shunt.

Case

A 25 year-old female G1P0 with a known past medical history of Chiari malformation type 1 with lumboperitoneal (LP) shunt, scoliosis s/p spinal fusion, mild aortic root dilation, and prior pericardial effusion who presented for evaluation and delivery planning.

Gestational Age	Prenatal Course
20 + 3 wks	Presents to obstetric cardiac clinic for prenatal evaluation. Cardiac MRI: trivial AI, 3.7 cm aortic root, 4 cm ascending aorta
26 + 2 wks	Given her combination of diagnoses, Cantu syndrome was suspected and genetic testing performed on the patient and fetus that demonstrated the ABCC9 gene mutation, consistent with Cantu syndrome
29 + 2 wks	ECHO demonstrates hyperdynamic LV EF 72%, aortic root and ascending aorta dilation, trivial AI
32 + 3 wks	Presents to obstetric cardiac clinic with tachycardia. Prescribed metoprolol succinate 12.5 mg daily.
35 + 1 wks	Presents with worsening dyspnea of exertion, musculoskeletal chest pain, and orthopnea. Repeat ECHO demonstrates aortic root dilation to 4 cm and ascending aorta dilation to 4.2 cm.
36+1 wks	Admitted with painful contractions for observation and rest.
36+6 wks	Patient had new onset vaginal bleeding. Decision made to proceed with Cesarean section for breech presentation. Patient underwent c-section under general anesthesia, as LP shunt and spinal fusion precluded neuraxial anesthesia.

Patient had an uncomplicated postpartum course and was discharged on PPD 6. Baby has a short stay in the NICU given prematurity and Cantu syndrome diagnosis but was also discharged to home as expected.

Anesthetic Considerations and Cantu Syndrome

Airway:

- Broad nasal bridge and large tongue impair mask ventilation
- Macroglossia, osteochondrodysplasia, and thickened calvarium may impair direct laryngoscopy, consider video laryngoscopy

Neuraxial Anesthesia:

- Wide vertebral bodies, scoliosis, and thickened calvarium leading to Chiari malformations may make neuraxial anesthesia difficult or unsafe

Cardiac abnormalities and Pregnancy:

- Increased blood volume can exacerbate valvular disorders, cardiomyopathy, and pericardial effusions – consider regular monitoring with transthoracic echocardiogram (TTE)

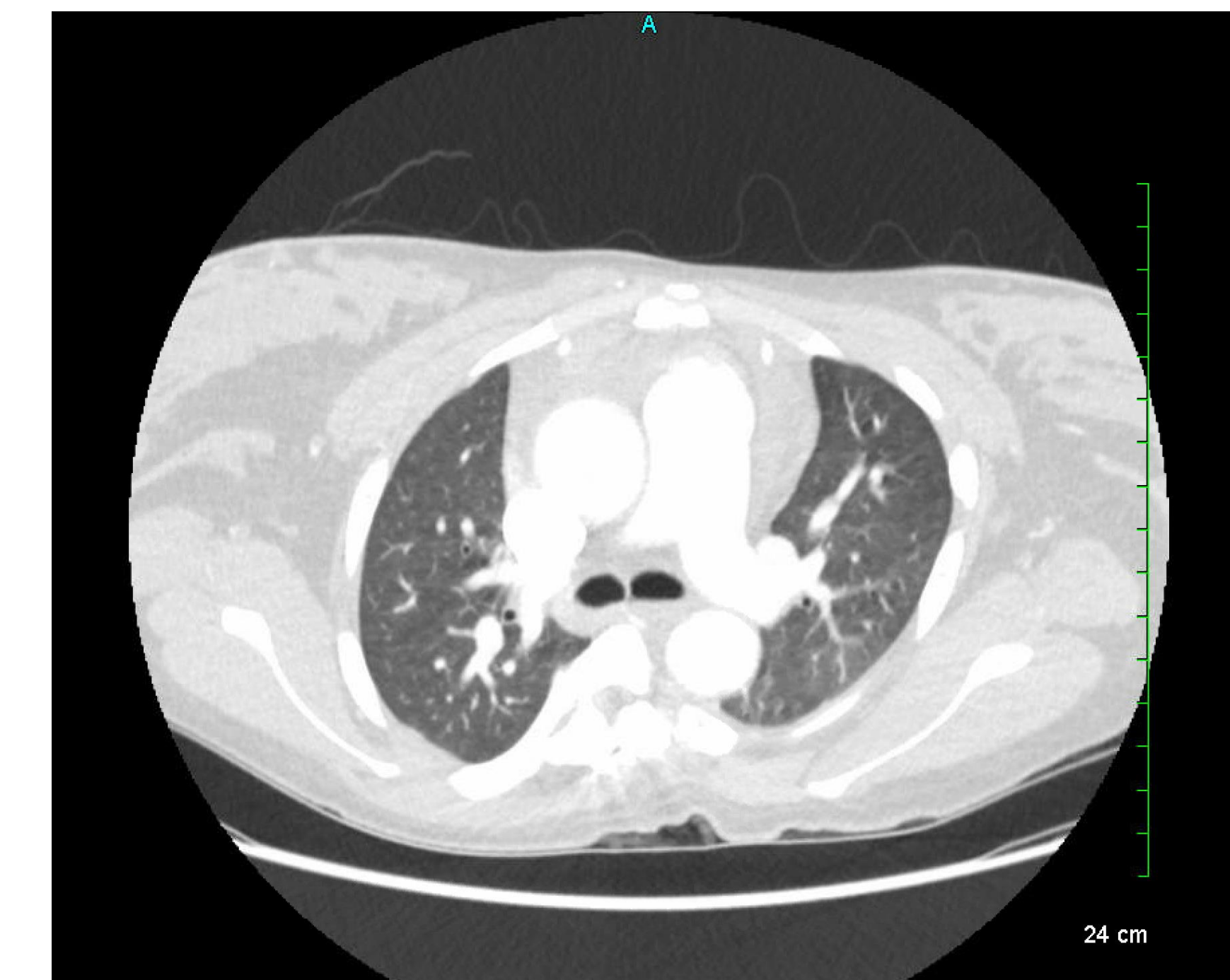


Figure 2. CTPA demonstrates dilated aortic root and pulmonary artery

References

- [1] Capdevila X., et al. "Continuous Peripheral Nerve Blocks in Hospital Wards after Orthopedic Surgery: A Multicenter Prospective Analysis of the Quality of Postoperative Analgesia and Complications in 1416 Patients". *Anesthesiology* 103 (5): 1035-1045.
- [2] Ilfeld B, "Continuous Peripheral Nerve Blocks: A Review of the Published Evidence." *Anesthesia and Analgesia* 113 (4): 904-915.