# Pulsatile Swelling of Umbilicus in a Cyanotic Neonate

Cantrell syndrome is a rare, usually lethal, congenital malformation [1]. In the complete form, five anomalies exist, namely a midline supra-umbilical abdominal wall defect, a sternal defect, an anterior diaphragmatic defect, a diaphragmatic pericardial defect and a congenital heart defect. However, the extent of individual defects and their combination varies considerably; broad spectrum of associated cardiac abnormalities have been reported in most cases. We describe a neonate presenting with a pulsatile umbilical swelling and cyanosis since birth, later confirmed to be due to Cantrell syndrome.

A full term male neonate with uneventful antenatal and perinatal course, born to a primigravida mother by normal vaginal delivery, was noted to have a pulsatile umbilical mass immediately after birth. Antenatal second trimester sonographic scans were reported normal, but detailed anomaly scan was not done. Baby was seen at our institute on day seven of life. He was feeding well, had a capillary filling time <3 second and normal urine output. Examination revealed tachycardia with a heart rate of 200 beats per minute, and central cyanosis (oxygen saturation 85% in room air). A peculiar mass arising from just above the umbilical stump, measuring 5×2×2 cm was noted. The mass enlarged with each cardiac systole (***Fig.* 1a**). It was covered with skin on its dorsal aspect but its ventral aspect was devoid of skin. On palpation, the structure had a forceful impulse and auscultation revealed a loud to-and-fro murmur over the mass. Cardiovascular examination revealed no evidence of heart failure, wide split second heart sound with a soft pulmonic component and a grade 3/6 ejection systolic murmur at left upper sternal border. A midline defect was palpable in the anterior abdominal wall.

Electrocardiogram revealed atrial flutter with ventricular rate of 200/min. Chest radiograph showed situs solitus, levocardia, normal sized heart and pulmonary oligemia. Ultrasound and Doppler evaluation of the mass revealed normal umbilical arteries, with a connection between umbilical vein and left ventricle through falciform ligament, suggestive of left ventricular diverticulum. Echocardiogram showed double outlet right ventricle with infundibular pulmonary stenosis. There was a tubular structure arising from apex of left ventricle which had a to-and-fro Doppler flow through it (***Fig.* 1b**). Computed tomography (CT) angiography revealed a dilated vascular channel beginning from the umbilical outpouching and travelling cranially within the anterior abdominal wall and along the falciform ligament to drain into the left ventricular apex through a 2 mm opening. There were multiple stenosis throughout its course, confirming the diagnosis of left ventricular diverticulum (***Fig.* 1c**). Defect in anterior diaphragm was present, but there was no associated sternal defect.

In view of atrial flutter, patient was started on propranolol and digoxin, and good heart rate control was achieved. After discussion with the cardiac surgical team, it was decided to close the left ventricular diverticulum, the cardiac lesion to be addressed later, since the oxygen saturation of the baby stayed above 85%. Left anterolateral thoracotomy was done and the fistulous tract arising from anterior most part of the left ventricular apex was identified. It was double clamped and divided and both ends were sutured. Defect in diaphragm was closed. The umbilical swelling was excised and the skin repaired. Patient had a smooth post-operative course and recovered well. The cardiac rhythm reverted to sinus rhythm on postoperative day 6. He was discharged on ninth post-operative day. Digoxin was stopped at six-week follow-up. Currently, at one-year follow up, baby is doing well, his oxygen saturation is 80% and he is in sinus rhythm. He is planned for Glenn surgery in view of non-committed muscular VSD, and is awaiting the same.

Only 250 cases of Cantrell syndrome have been reported in the literature [2]. It has high morbidity and mortality, with more than half of patients dying, many despite surgery [3]. Abnormal migration of the splanchnic and somatic mesoderm (which affects the development of the heart and the major vessels) with premature breakage of the chorion or vitelline sac at about day 14 to 18 of gestation, may lead to a mid-line defect [4] Congenital cardiac malformations are associated in majority,

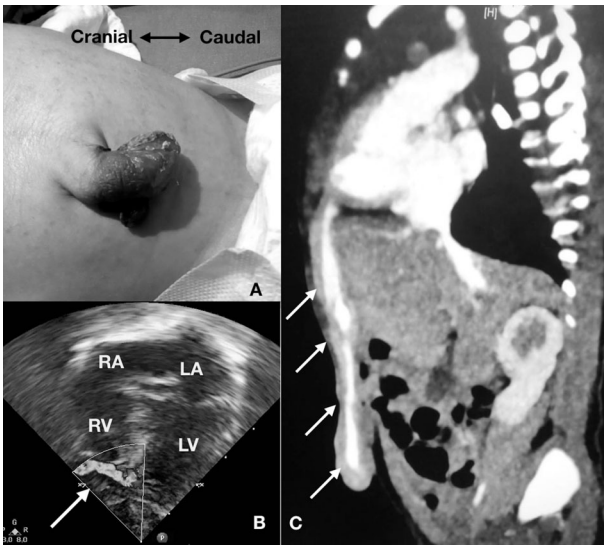


**Fig. 1** *(a) Clinical photograph of umbilical swelling; (b) echocardiogram in four chamber view showing a vascular channel (white arrow) arising from left ventricle apex; (c) CT angiographic section in sagittal view showing the abnormal vascular channel (white arrows) arising from left ventricle apex.*

ventricular septal defect is the commonest abnormality. Association with double outlet right ventricle has also been previously reported [5].

Over 70% of patients with left ventricular diverticulum have Cantrell syndrome. The diverticulum originates from the left ventricular apex in these cases and may be associated with umbilical hernia and complex cardiac abnormalities. Ventricular aneurysm must be differentiated from diverticulum. A narrow mouth and synchronous contractility characterize a diverticulum. On the other hand, aneurysms show akinesia or paradoxic contractility of the outpouching, which is asynchronous with the rest of heart.

Early surgical repair is indicated in cases of left ventricular diverticulum, as it may rupture spontaneously, thrombose or produce arrhythmias. It is generally recommended that the midline thoraco-abdominal defect is treated first and heart defects be corrected later [6]. We present this case in view of the interesting presentation in a neonate with a pulsatile umbilical swelling and cyanosis, and a good outcome after surgery.

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**REFERENCES**

1. Cantrell JR, Haller JA, Ravitch MM. A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart. Surg Gynecol Obstet. 1958;107:602-14.
2. Jnah AJ, Newberry DM, England A. Pentalogy of Cantrell: Case report with review of the literature. Adv Neonatal Care. 2015;15:261-8.
3. O'gorman CS, Tortoriello TA, McMahon CJ. Outcome of children with pentalogy of Cantrell following cardiac surgery. Pediatr Cardiol. 2009;30:426-30.
4. Amato JJ, Douglas WI, Desai U, Burke S. Ectopia cordis. Chest Surg Clin North Am. 2000;10:297-316.
5. Singh N, Bera ML, Sachdev MS, Aggarwal N, Joshi R, Kohli V. Pentalogy of Cantrell with left ventricular diverticulum: A case report and review of literature. Congenital Heart Dis. 2010;5:454-7.
6. Williams AP, Marayati R, Beierle EA. Pentalogy of Cantrell. Semin Pediatr Surg. 2019;28:106-10.

# Mechanical Thrombectomy for Cerebral Venous Sinus Thrombosis in a Neonate

Cerebral venous sinus thrombosis has a reported incidence of 0.35-0.67 per 100000 children per year, and about 40% of cases occur during the neonatal period [1]. In the pediatric population, standard choice of treatment is the use of low molecular weight heparin (LMWH). The indication criteria and the role of mechanical thrombectomy and other interventional procedures in infants with cerebral venous thrombosis is unknown [2,5]. Even in neonates treated with LMWH, the incidence of neurological disability is unfavorably high,especially in those with multi-sinus thrombosis [1]. We report a neonate with this disorder managed with mechanical thrombectomy.

A 10-day-old term male neonate presented to the pediatric emergency department with partial seizure of the upper extremities. The infant was born after an uncomplicated pregnancy followed by a normal spontaneous vaginal delivery and was discharged home from neonatal nursery after 72 hours, with no need of intervention and medication. At presentation, the infant was afebrile, apathetic, with feeding difficulties, and had mild tachycardia and delayed capillary refill time. A weight loss of 15% compared to the discharge weight was noted. The clinical state was evaluated as dehydration, and intravenous rehydration was started. Two hours after admission, myoclonic seizures of upper extremities occurred, along with multiple apneic spells reappeared. Anticonvulsant treatment with intravenous phenobarbital was started. Laboratory examinations (blood count, plasma minerals and serum biochemistry, C-reactive protein, procalcitonin, coagulation profile) and lumbar puncture results were unremarkable, except for lactate concentration (4.75 mmol/L), hematocrit level (61%) and hemoglobin concentration (20 g/dL). Magnetic resonance imaging (MRI) with consecutive time-of-flight (TOF) venography and contrast enhanced T1WI revealed cerebral venous thrombosis. Superior sagittal sinus, right transverse sinus, straight sinus, vein of Galen and internal cerebral veins thrombosed, along with hemorrhage from right choroid plexus, and bilateral thalamic vasogenic edema. After multi-specialty consultation, mechanical thrombectomy was planned, in view of the presence of multi-sinus thrombosis with thalamic edema and signs of neurologic deterioration with acute repetitive seizures.

After obtaining informed consent from the baby's mother, the procedure was performed under general anesthesia with ultrasound control. The right internal jugular vein was punctured and a 3F introducer (IVA, BALT) was placed by the Seldinger technique. The microcatheter (Orion, Medtronic) was navigated *via* micro-guide wires Hybrid .008" and Hybrid.1214DA (BALT) into the straight sinus as well as into the superior sagittal sinus directly without the use of a guide catheter. Mechanical thrombectomy was performed *via* a Solitaire platinum 6ã40 (Medtronic) three times per sinus. Hemostasis in the puncture site was achieved by compression with usage of HemCon Patch.