

Pentalogy of Cantrell and anaesthesia: a case report

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Pentalogy of Cantrell, a rare congenital thoracoabdominal disruption, consists of a supraumbilical abdominal wall defect, lower sternal defect, agenesis of the anterior portion of the diaphragm, an absence of the diaphragmatic part of the pericardium, and a malformation of the heart, typically as an ectopia cordis. This case report presents a female neonate, 35 weeks post-conception, weighing 2080 g who presented on day four of life for anaesthesia. She had the five anatomical defects characteristic of Pentalogy of Cantrell.

Keywords: abdominal wall defects, anaesthesia, congenital heart disease, ectopia cordis, extrathoracic heart, Pentalogy of Cantrell, perioperative management, transfer difficulties in South Africa

Case report

A female neonate, day two of life, born at 35 weeks' gestation at a peripheral hospital, was transferred to Chris Hani Baragwanath Academic Hospital (CHBAH) with a provisional diagnosis of Pentalogy of Cantrell (POC). APGARs were 7 and 9 at 1 and 5 min respectively. She weighed 2080 g. The mother was a 20-year-old primigravida with no medical history. She tested negative for the human immunodeficiency virus. There was no positive family history of congenital cardiac abnormalities. The transfer to the tertiary facility took 5 h. On arrival at CHBAH neonatal ICU she was haemodynamically stable on nasal prong oxygen at 2 l/minute which was initiated immediately after birth. Her cardiac defect had been covered with a transparent plastic silo for the transfer in an attempt to prevent sepsis. She was hypoglycaemic, which was corrected.

On examination she appeared dysmorphic with micrognathia, microphthalmia, a flattened nasal bridge and hypertelorism. There was a large defect (6 × 4 cm) in the thoracoabdominal wall with ectopia cordis (EC) (heart and great vessels exposed) and absent pericardium. She had a sternal cleft, a small omphalocele and an anterior diaphragmatic defect (Figure 1). No pulmonary issues were noted but she appeared to have ambiguous genitalia; all her limbs appeared normal. She was diagnosed as a Class 1 POC according to Toyama's definition. She was passing urine and meconium.

A chest X-ray (Figure 2) revealed an abnormal position of her heart, reduced pulmonary vasculature and an abdominal mass centrally in keeping with the omphalocele. Echocardiography demonstrated an atrial septal defect (ASD) measuring 5.6 mm and a small patent ductus arteriosus (PDA). Basic blood investigations were normal and genetic testing results were unavailable.

A multidisciplinary discussion involved Cardiothoracic Surgery, Paediatric Surgery, Anaesthesia and ICU. Since CHBAH has no cardiothoracic facilities, in order to prevent delay with another transfer it was decided that the cardiothoracic surgeons would come to CHBAH to perform the procedure. This is common practice in our facility with other cardiothoracic procedures such as our PDA ligations. Written informed consent was obtained from the parents. On day four of life she was scheduled for a repair or coverage of her EC and abdominal defect.

She arrived in theatre comfortable on nasal prong oxygen. She was physiologically stable (blood pressure 75/46 (56); heart rate 154 bpm with oxygen saturations (sats) of 98%, apyrexial) and cried upon stimulation. Air entry was clear bilaterally with no additional sounds. She had a 24 G intravenous (IV) line in situ. Blood results reflected a normal full blood count (haemoglobin 12.3 g/dl) and electrolytes. She was receiving intravenous antibiotics, which had been initiated on admission to ICU in view of her high risk for sepsis.

Standard ASA monitors were applied, a fluid warmer attached to the IV line and Bair Hugger™ (3M, St Paul, MN, USA) placed. After pre-oxygenation, an elective sequence induction was performed, bag-mask ventilation was confirmed, and she was paralysed. She had a difficult airway and desaturated with the first attempt at intubation with no associated bradycardia. Cricoid pressure was then applied and external laryngeal manipulation to the left allowed vocal cord visualisation. A size 3.0 uncuffed endotracheal tube was placed and she was ventilated with pressure control ventilation (PCV) and maintained with sevoflurane in an oxygen and air mixture. She received fentanyl for analgesia and invasive monitoring was placed. Surgery lasted 137 min. She did require a blood transfusion and despite using multiple warming devices hypothermia to 33°C occurred. During manipulation and compression of the heart on the left side there was a marked reduction in cardiac output (CO), which self-corrected when the manipulation stopped. She suffered no cardiac arrest. The surgeons fashioned a patch to fit the shape of the heart as a



Figure 1: Complete Pentalogy of Cantrell.



Figure 2: Plain thoracoabdominal film (rotated).

rudimentary pericardium and a GoreTex® soft tissue patch was used to cover the abdominal defect. To reduce the risk of contamination, a sterile 500 ml saline bag was sutured to cover both patches (Figure 3). After surgery, tracheal intubation was maintained and the infant was warmed to 35.5° C before being transferred haemodynamically stable (blood pressure 83/68, mean arterial pressure 73, heart rate 125, sats 93%) with a normal blood gas to the neonatal ICU.



Figure 3: A sterile saline bag covering both patches.

Postoperatively, after 12 h of stability, she developed signs of sepsis and continued to deteriorate requiring inotropic support. A methicillin-resistant *Staphylococcus aureus* sensitive to vancomycin was documented on blood cultures. Despite maximal therapy the baby died 48 h after surgery.

Discussion

POC, first described by Cantrell *et al.* in 1958, is a rare congenital anomaly involving a defect in the thoracoabdominal wall.^{1,2} It is a syndrome of variable expression. In its complete form, it is characterised by the following five defects:

- (1) supraumbilical abdominal wall defect with omphalocele;
- (2) lower sternal defect;
- (3) ectopia cordis;
- (4) anterior diaphragmatic defect;
- (5) diaphragmatic pericardial defect and intracardiac abnormalities.³

The intracardiac associations are typically ventricular septal defects, atrial septal defects, pulmonary stenosis, Tetralogy of Fallot and left ventricular diverticulae.^{1,4,5} By identifying the typical features on ultrasonography, POC can usually be diagnosed at the end of the first trimester.³

The degree of severity varies and was classified by Toyama in 1972 based on a review of 61 cases of POC. He divided the pentalogy into three classes based on the spectrum of expression:

Class 1—complete form of the syndrome, having all five defects (definitive diagnosis);

Class 2—four defects including ventral wall and intracardiac abnormalities (probable diagnosis);

Class 3—incomplete form having various combination of defects but with sternal abnormality (incomplete expression).⁶

The prevalence of POC is around 5.5 per 1 million live births⁷ and occurs sporadically^{7,8} although there is evidence for X-linked dominant inheritance.² It is due to a developmental abnormality at about 3–4 weeks post-conception in the ventral mesoderm.^{4,9}

A multidisciplinary approach is essential in planning the management of these cases.^{5,7} Should they survive, these cases require multiple surgeries. The aim of surgery is to facilitate soft tissue cover of the exposed organs (the heart and abdominal viscera) and to reduce them into their respective cavities. These cases also require a repair or palliation of the intracardiac lesions and reconstruction of the abdominal and chest defects after appropriate growth of the thoracic cavity and lungs.¹⁰ Corrective and palliative cardiothoracic surgery has been provided with success for these cases.⁷

Anaesthetic considerations

Airway

The airway may potentially be difficult, especially with other associated midline defects² and possible orofacial abnormalities.²

Respiratory

Pulmonary hypoplasia or agenesis of the left lung, with or without a diaphragmatic hernia, may be present.⁵ The resultant mechanical dysfunction of the lungs may be worsened by external pressure and may worsen the haemodynamic effects of PCV. One should ensure no compression of the lower part of the sternum or abdomen when positioning the patient⁵ and maintain low pulmonary inflation pressures to reduce barotrauma and potential pneumothorax.²

Cardiovascular

In the severest form, EC may be present with varying intracardiac lesions. The pericardium is absent in 75% of cases.¹¹ Haemodynamic changes may be due to either compression of the exposed heart or overlapping complications of potential associated cardiac malformations and in some cases a combination.⁴

Abdominal

Omphalocele in 63% of cases, polysplenia and gallbladder agenesis can be present.¹¹ Monitoring for acute respiratory and cardiac failure is essential because of the potential for raised intra-abdominal pressure with reduction of the omphalocele⁴ and resultant herniation of the reduced abdominal contents through the diaphragmatic hernia.² High intra-abdominal pressures may also compromise renal perfusion and may splint the diaphragm further, compromising respiratory and cardiac function.²

There is a risk of hypothermia and fluid/electrolyte losses due to exposure of the heart and viscera.⁴

Other anomalies may include: hydrocephalus¹¹ or anencephaly,² cleft lip/palate¹¹ with resultant potentially difficult airway, renal or skeletal abnormalities and sternal defects.¹¹

The postoperative period remains critical. Postoperative monitoring in a paediatric ICU is essential, using adequate analgesia and where possible early weaning from the ventilator. Postoperative apnoea is a risk and prophylactic intravenous caffeine is recommended in premature neonates, but one should be aware of the potential side effects of its use such as increased work of breathing, excitability and tachycardia.⁵

A South African perspective

A similar case has previously been published by Dr A. Bösenberg in 2004² but the patient, who also died, never made it to theatre. In both cases the treating teams were faced with delays due to lack of specialised facilities at the peripheral hospitals and transport issues. In our case specifically it was an unexpected presentation as it was not diagnosed prenatally on ultrasound, which would have facilitated planning of her delivery in a tertiary institution. In South Africa, it is common practice for children to be referred from lower-level facilities to higher-level facilities to access resources, specialist knowledge and skills for diagnosis and treatment.¹² Our South African experience shows the limited capacity to provide adequate care to complicated cases that present in the peripheral health service, as is evidenced by up to 34% mortality for these referrals within the first 24 h of admission to a tertiary facility.¹³ There is a reliance on a referral system in the current South African public health care system due to centralisation for the provision of specialist paediatric and intensive care in tertiary institutions.¹² Due to the vast resource constraints and distances to be travelled in some regions of the country, appropriate and timely referral is quite difficult.¹⁴

In conclusion, these patients generally have a poor prognosis depending on the severity of the cardiac and associated malformations.² The specific treatment strategy will vary between patients depending on the size and type of abdominal wall defect, the specific cardiac anomalies that are present, and the particular type of EC. It is essential for the anaesthetic team to understand the planned procedure as this will help to anticipate and manage the potential complications intraoperatively. In our case a staged repair of the defects associated with POC was planned. The initial operation provided separation of the peritoneal and pericardial cavities, and coverage of the midline defect and of the omphalocele¹⁵ but unfortunately the infant died due to sepsis.

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References

- Cantrell JR, Haller JA, Ravitch MM. A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. *Surg Gynecol Obstet.* 1958 Nov;107(5):602–14. PubMed PMID: 13592660.
- Bösenberg A. Anaesthesia and pentalogy of cantrell. *SAJAA.* 2004 Oct;10(4): 5–7.
- Mohamed ASK. Pentalogy of cantrell [Internet]. [Cited 2013 July 02–09]. Pentalogy of Cantrell © Anwer Sadat Kithir Mohamed www.TheFetus.net. [cited Dec 6, 2016]. Available from: <https://www.sonoworld.com/fetus/page.aspx?id=3339>.
- Suehiro K, Okutani R, Ogawa S, et al. Perioperative management of a neonate with Cantrell syndrome. *J Anesth.* 2009;23:572–5. doi:10.1007/s00540-009-0785-9. PubMed PMID: 19921368.
- Laloyaux P, Veyckemans F, Van Dyck M. Anaesthetic management of a prematurely born infant with Cantrell's pentalogy. *Pediatr Anaesth.* 1998;8 :163–6. PubMed PMID: 9549746.
- Toyama WM. Combined congenital defects of the anterior abdominal wall, sternum, diaphragm, pericardium, and heart: a case report and review of the syndrome. *Pediatrics.* 1972 Nov;50(5):778–92. PubMed PMID: 4263752.
- Sowande OA, Anyanwu LJC, Talabi AO, et al. Pentalogy of Cantrell: A Report of Three Cases. *J Surg Tech Case Rep.* 2010 Jan–Jun;2 (1):20–3. doi:10.4103/2006-8808.63717. PubMed PMID: PMC3214484
- Polat I, Gül A, Aslan H, et al. Prenatal diagnosis of pentalogy of Cantrell in three cases, two with craniorachischisis. *J Clin Ultrasound.* 2005;33:308–11. doi:10.1002/jcu.20134. PubMed PMID: 16134161
- Duhamel B. Embryology of exomphalos and allied malformations. *Arch Dis Child.* 1963;38:142–7. PubMed PMID: 21032411.
- Lack V, Oetzmann von Sochaczewski AC, Naidoo K, et al. Pentalogy of Cantrell with thoracoabdominal ectopia cordis: attempted surgical correction and review of recent literature to aid prognostication prior to surgery. *J Ped Surg Case Reports* 3. 2015;3:476–80.
- Chandran S, Ari D. Pentalogy of Cantrell: an extremely rare congenital anomaly. *J Clin Neonatol.* 2013;2 (2) April–June:95–7. doi:10.4103/2249-4847.116410. PubMed PMID: 24049753.
- Royal C, McKerrow NH. A retrospective review of the transfer of critically ill children to tertiary care in KwaZulu-Natal Province, South Africa. *South Afr J Child Health.* 2015 Nov;9(4):112–8. doi:10.7196/SAJCH.2015.v9i4.913
- Stephen CR, Bamford LJ, Patrick ME, et al. editors. *Saving Children 2009: five years of Data. A sixth survey of child healthcare in South Africa.* Pretoria: Tshepesa Press, MRC, CDC; 2009.
- Scribante J, Bhagwanjee S. National audit of critical care resources in South Africa—transfer of critically ill patients. *South Afr Med J.* 2007;97(12):1323–6.
- Genetic and Rare Disease Information Center. Pentalogy of Cantrell [Internet]. [updated 2009 May 27; cited 2017 Jan 2]. Available from: <https://rarediseases.org/rare-diseases/pentalogy-of-cantrell>.

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