Eur J Pediatr (2008) 167:29–35
DOI 10.1007/s00431-007-0578-9

REVIEW

Pentalogy of Cantrell: two patients and a review to determine prognostic factors for optimal approach

Jeroen H. L. van Hoorn · Rob M. J. Moonen · Clément J. R. Huysentruyt · L. W. Ernest van Heurn · Jos P. M. Offermans · A. L. M. Twan Mulder

Received: 7 April 2007 / Accepted: 12 July 2007 / Published online: 4 August 2007

Abstract Two patients with incomplete pentalogy of Cantrell are described. The first was a girl with a large omphalocele with evisceration of the heart, liver and intestines with an intact sternum. Echocardiography showed profound intracardiac defects. The girl died 33 h after birth. The second patient was a female fetus with ectopia cordis (EC) without intracardiac anomalies; a large omphalocele with evisceration of the heart, stomach, spleen and liver; a hypoplastic sternum and rib cage; and a scoliosis. The pregnancy was terminated. A review of patients described in the literature is presented with the intention of finding prognostic factors for an optimal approach to patients with the pentalogy of Cantrell. In conclusion the prognosis seems to be poorer in patients with the complete form of pentalogy of Cantrell, EC, and patients with associated anomalies. Intracardial defects do not seem to be a prognostic factor.

Keywords Ectopia cordis · Pentalogy of Cantrell · Abdominal wall defect

Statement of financial support: no financial assistance was received.

J. H. L. van Hoorn · R. M. J. Moonen · A. L. M. T. Mulder (✉)
Department of Pediatrics, University Hospital Maastricht,
P.O. Box 5800, 6202 AZ Maastricht, The Netherlands
e-mail: amul@paed.azm.nl

C. J. R. Huysentruyt
Department of Pathology, University Hospital Maastricht,
P.O. Box 5800, 6202 AZ Maastricht, The Netherlands

L. W. E. van Heurn
Department of Surgery, University Hospital Maastricht,
P.O. Box 5800, 6202 AZ Maastricht, The Netherlands

J. P. M. Offermans
Department of Obstetrics and Gynaecology,
University Hospital Maastricht,
P.O. Box 5800, 6202 AZ Maastricht, The Netherlands

Abbreviations

ASD	atrial septal defect
EC	ectopia cordis
MRI	magnetic resonance imaging
VSD	ventricular septal defect

Introduction

The pentalogy of Cantrell was first described in 1958 [10]. The hallmark of this syndrome is an omphalocele associated with ectopia cordis (EC). The full spectrum consists of five anomalies: a deficiency of the anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, various congenital intracardiac abnormalities, and a defect of the lower sternum. Only a few patients with the full spectrum of the pentalogy have been described. We present two patients with incomplete pentalogy of Cantrell. We reviewed the literature to find prognostic factors that may help to assess the best multidisciplinary approach in prenatal counselling and in postnatal therapy in patients with the pentalogy of Cantrell.

Case reports

Patient 1

A prenatal ultrasound in a 26-year-old G1P0 showed a fetus with bilateral hydrothorax, EC with a ventricular septal defect (VSD), and a large omphalocele with evisceration of the heart and the liver. The diagnosis of pentalogy of Cantrell and the prognosis were discussed with the parents. The prenatal medical team together with the parents decided to continue the pregnancy. At 39 weeks and

1 day of gestational age, a girl was delivered by primary cesarean section with Apgar scores of 6 and 8 at 5 and 10 min, respectively. Birth weight was 2,310 g (<p 2,3). There was a large omphalocele with evisceration of the heart, liver and intestines (Fig. 1); on palpation the sternum was intact. Echocardiography showed tetralogy of Fallot with a VSD, pulmonary valve stenosis and an aberrant aortic valve, a large atrial septal defect (ASD), and signs of pulmonary hypertension. The girl was intubated 30 min after birth. Due to progressive respiratory distress, conventional mechanical ventilation was switched to high-frequency oscillation. Endotracheal instillation of surfactant and evacuation of 45 ml pleural fluid were performed without any clinical improvement. Treatment with inhaled nitric oxide, inotropic support of the heart and systemic blood pressure, and prostaglandin E1 (Prostin VR Paediatric) to preserve the ductus-dependent circulation were started. Despite this treatment, the child remained hypotensive with oxygen saturation levels between 50 and 60%. The girl died 33 h after birth. The parents refused autopsy.

Patient 2

A prenatal ultrasound in a 19-year-old G1P0 showed a fetus with EC with a VSD; a large omphalocele with evisceration of the heart, stomach, spleen, and liver; and a scoliosis. After discussing the diagnosis of pentalogy of Cantrell and related prognosis with the parents, the pregnancy was terminated at 23 weeks and 4 days of gestational age.

Post-mortem examination showed a female fetus presented with a large omphalocele with evisceration of the liver, spleen, and a major part of the gastro-intestinal tract (Fig. 2). The heart was situated directly under the skin, not protected by the ribs or the hypoplastic sternum. The anterior diaphragm was absent and a peritoneal-pericardial connection was found. Furthermore the fetus was characterized by a low implant of the left ear, a severe scoliosis, and a hypoplastic right ribcage with both lungs positioned in the left ribcage. Because of its small size, the heart was examined under the dissecting microscope, but a VSD or another intracardial defect could not be found.

Histopathological examination showed dysmaturity of the lungs due to the intrathoracal malpositioning. The other organs showed no major abnormalities on microscopy.

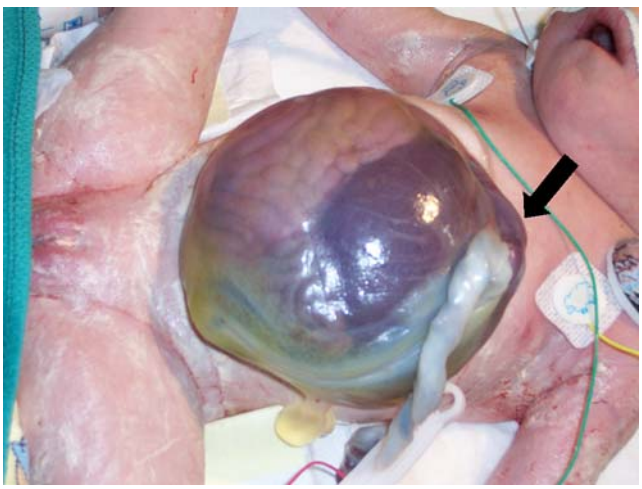

Fig. 1 Patient 1 with a large omphalocele with evisceration of the heart (*arrow*), liver and intestines

Discussion

The pentalogy of Cantrell is a rare syndrome with an estimated incidence of 5.5 per 1 million live births [11]. It is described as a deficiency of the anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, various congenital intracardiac abnormalities, and a defect of the lower sternum. The pathogenesis of pentalogy of Cantrell has not been fully

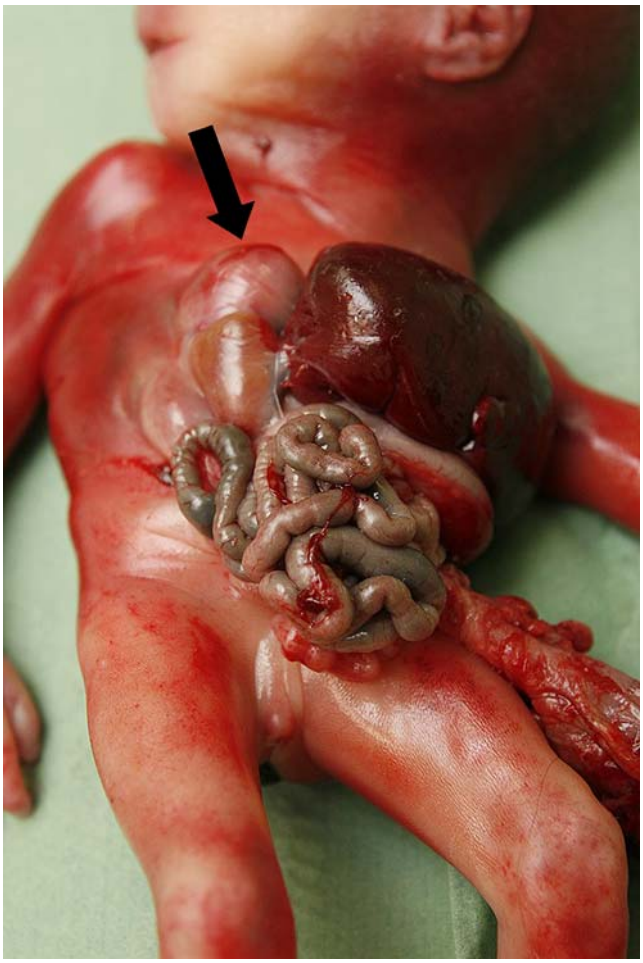

Fig. 2 Post-mortem examination of patient 2 showed a large omphalocele with evisceration of the liver, spleen and a major part of the gastro-intestinal tract. The *arrow* indicates the heart covered with skin

Table 1 Review of patients with pentalogy of Cantrell (complete and incomplete form) with cardial defects, associated anomalies, and outcome

Reference	Form	Cardial defect	Associated anomalies	Survival
Baker et al. 1984 [5]	CF	EC	Cloacal extrophy, genitourinary and spine anomalies	No
Soper et al. 1986 [38]	CF	EC, VSD	Occipital encephalocele 47,XX +18	No
Bick et al. 1988 [7]	CF	EC	Occipital encephalocele 47,XX +18, abnormally lobated small lungs, horseshoe kidney	No
Fox et al. 1988 [18]	CF	EC, single ventricle and atrium, bicuspid ventricular outflow valve, malpositioned right aortic arch	Bilateral clubfeet, spina bifida, hydrocephalus, abnormal ears, horseshoe kidneys: trisomy 18	No
Carmi and Boughman 1992 [11]	CF	EC, VSD, D-transposition of great vessels, pulmonary atresia, hepatic venous connection, common pulmonary vein	Large left cleft lip and palate	No
	CF	EC, diverticulae of right and left ventricle	None	Yes
	CF	EC, TOF	Bilateral cleft lip and palate, single-lobed left lung, intralobar pulmonary sequestration	No
	CF	TOF, ASD	None	No
	CF	TOF, pulmonary atresia	Right cleft lip, small ears, dysplastic toe nails	No
Martin et al. 1992 [27]	CF	EC, hypoplastic left ventricle and atrium, dilated right ventricle, pulmonary valve and artery	None	No
	CF	ASD, VSD	None	No
Egan et al. 1993 [17]	IF	EC, hypoplastic left ventricle, single pulmonary vein	Sirenomelia	No
Abdallah et al. 1993 [1]	CF (?)	EC, TOF	None	Yes
Bogers et al. 1993 [9]	CF	EC, VSD, left and right ventricular diverticulum	None	Yes
Denath et al. 1994 [15]	IF	EC	Exencephaly, pulmonary hypoplasia	No
Siles et al. 1996 [36]	IF		None	Yes
	IF		None	Yes
	IF	VSD, double outlet right ventricle, bilateral superior venae cavae, pulmonary stenosis	None	Yes
Chen et al. 1996 [13]	CF (?)		None	Yes
Hornberger et al. 1996 [21]	?		None	No
	?		None	No
Liang et al. 1997 [25]	IF	EC	None	No
Vazquez-Jimenez et al. 1998 [42]	IF	ASD, VSD, LVD, left superior vena cava without connection to the right system	Short, flat nose	Yes
Hsieh et al. 1998 [22]	CF	EC	Cystic hygroma	No
	CF	EC	Cystic hygroma	No
Laloyaux et al. 1998 [24]	CF	VSD, ASD, tricuspid atresia, pulmonary stenosis	None	Yes
Song et al. 2000 [37]	IF	EC, single ventricle with double outlet, pulmonary atresia, tricuspid atresia	None	No
Morales et al. 2000 [29]	IF	EC, VSD, LVD, dextrocardia, double outlet right ventricle, right ventricle outflow tract obstruction, dextrocardia, VSD, double outlet right ventricle, pulmonary stenosis	Cleft palate, large encephalocele, hydrocephalus	Yes
	IF	TOF	None	Yes
	IF		None	Yes
Alayunt et al. 2001 [2]	CF (?)	VSD, LVD, ASD, TOF	None	Yes

Table 1 (continued)

Reference	Form	Cardial defect	Associated anomalies	Survival
Spencer et al. 2002 [39]	CF (conjoined twin)	EC, single anomalous multiventricular heart with ventricular septal defects, single common atria with three atrioventricular openings, anomalous systemic and pulmonary venous drainage; one twin: severe pulmonary stenosis; other twin: absent ductus ateriosus; common atria bilateral superior venae cavae; tricuspid, mitral and pulmonary valve aplasia; malrotation of the great vessels; aorticopulmonary communication	Thoracopagus twins	No
	CF (conjoined twin)	ASD	Omphalopagus twins	No
Halbertsma et al. 2002 [20]	IF	LVD, ASD, VSD, anomalous venous pulmonary return	None	Yes
Nanda et al. 2003 [30]	CF	EC, VSD	Kyphoscoliosis, club foot	No
	CF	EC, VSD	None	No
Onderoglu et al. 2003 [32]	CF	EC	Trisomy 21, pulmonary and extremity anomalies	No
Oka et al. 2003 [31]	CF	EC, PDA, LVD, tricuspid atresia, pulmonary stenosis, hypoplastic pulmonary arteries	None	Yes
Bittmann et al. 2004 [8]	CF	Small right ventricle, VSD, ASD	Gallbladder agenesis, polysplenia, segmentation defect of the lungs	No
Uygur et al. 2004 [41]	IF	EC Patent foramen ovale PDA	Left clubfoot, hypodactyly right hand, absent third finger of the right hand, absent left tibia and right radius	No
Aslan et al. 2004 [4]	IF	EC	Bilateral undescended testes, scoliosis, adherence between left upper limb and trunk, adrenohepatic fusion, anterior thoracic myeloschisis, multiple accessory spleens, renal agenesis	No
	IF	EC		No
Correa-Rivas et al. 2004 [14]	CF	EC, ASD, PDA	Bilateral cleft lip and palate, bilateral pulmonary hypoplasia	No
Polat et al. 2005 [34]	CF	EC	Craniorachischisis, bilateral clubfoot	No
	CF	EC	Craniorachischisis, bilateral clubfoot and clubhand	No
	CF	EC	None	No
Marijon et al. 2006 [26]	IF	LVD, VSD	None	Yes
Bhat et al. 2006 [6]	IF	Dextrocardia, ASD	None	Yes
Araujo Junior et al. 2006 [3]	CF	EC, VSD	None	No
Knirsch et al. 2006 [23]	IF	Mesocardia, VSD, ASD, LVD	None	Yes
Chen et al. 2006 [12]	CF	EC, VSD	Scoliosis, hypoplasia of the right upper limb, ectrodactyly of the right hand and foot	No
Rashid et al. 2007 [35]	CF	EC	Encephalocele, club foot	No
Desselle et al. 2007 [16]	CF	EC, TOF	Non-rotation of the midgut, accessory spleen	Yes
Grethel et al. 2007 [19]	IF	Ventricular aneurysm	Morgagni hernia	Yes

Table 1 (continued)

Reference	Form	Cardial defect	Associated anomalies	Survival
McMahon et al. 2007 [28]	IF	EC, TOF, VSD, hypoplastic pulmonary valve	None	?
	IF	EC, VSD	None	?
Our patients	IF	EC, TOF, ASD aberrant aortic valve	None	No
	IF	EC	Low implant of left ear, hypoplastic right rib cage,scoliosis	No

CF Complete form, *IF* incomplete form, *EC* ectopia cordis, *VSD* ventricular septal defect, *ASD* atrial septal defect, *TOF* tetralogy of Fallot, *LVD* left ventricular diverticulum, *PDA* patent ductus arteriosus

elucidated. Cantrell et al. [10] suggested an embryologic developmental failure of a segment of the lateral mesoderm around gestational age 14–18 days. Consequently, the transverse septum of the diaphragm does not develop, and the paired mesodermal folds of the upper abdomen do not migrate ventromedially. Organs may eviscerate through the resulting sternal and abdominal wall defects. EC itself is characterized by complete or partial displacement of the heart outside the body. Cervical, cervicothoracic, thoracic, and thoracoabdominal types of EC have been described [29].

Intracardiac anomalies are described in the pentalogy of Cantrell including VSD (100%), ASD (53%), tetralogy of Fallot (20%), and ventricular diverticulum (20%) [10]. Various other associated anomalies have been reported and include craniofacial and central nervous system anomalies such as cleft lip and/or palate, encephalocele, hydrocephalus, and craniorachischisis [14, 29, 34]; limb defects such as clubfoot, absence of tibia or radius, and hypodactyly [33, 41]; and abdominal organ defects such as galbladder agenesis and polysplenia [8]. Often the spectrum of the original pentalogy of Cantrell is not complete. Toyama [40] suggested the following classification of the pentalogy of Cantrell: class 1, definite diagnosis, with all five defects present; class 2, probable diagnosis, with four defects present, including intracardiac and ventral wall abnormalities; and class 3, incomplete expression, with various combinations of defects present, including a sternal abnormality. In our first patient, the sternum was intact, and in addition to the large omphalocele, there were diaphragmatic and intracardiac defects. The second patient had a sternal defect with associated anomalies such as a low implant of the left ear, a hypoplastic right rib cage and a scoliosis. There were no intracardiac anomalies. We considered both patients to be incomplete forms of the pentalogy of Cantrell.

With prenatal ultrasonography, the pentalogy of Cantrell usually can be diagnosed in the first trimester of pregnancy [25]. In a fetus with omphalocele, pentalogy of Cantrell should be ruled out. If pericardial effusion can be seen, associated anterior diaphragmatic hernia and diaphragmatic pericardial defects may be suspected, and specific and detailed search for the features of the pentalogy of Cantrell, as described above, should be done [36]. Use of prenatal magnetic resonance imaging (MRI) may enhance the visualization of the fetal anomalies [28].

After birth, echocardiography is essential for diagnosis of associated cardiac anomalies. Other features of the pentalogy of Cantrell and known associated anomalies can be diagnosed by conventional radiography or sonography. Nevertheless, small defects of the diaphragm and pericardium can be extremely difficult to diagnose accurately. In these patients and in cases of possible surgical intervention, MRI might be useful [31, 37].

The treatment of the pentalogy of Cantrell consists of corrective or palliative cardiovascular surgery, correction of ventral hernia and diaphragmatic defects and correction of associated anomalies. The best treatment strategy depends on the size of the abdominal wall defect, the associated heart anomalies, and the type of EC.

To find prognostic factors that might help to determine the best strategy in patients with the pentalogy of Cantrell we performed a literature search of patients with pentalogy of Cantrell described in the English literature over the last 20 years. The results are shown in Table 1. An overview of patients described earlier was made by Toyama et al. in 1972 [40]. Our search on Pubmed using the search terms “pentalogy of Cantrell” and “Cantrell’s syndrome” yielded 58 patients with pentalogy of Cantrell between 1987 and April 2007. Thirty-three patients were described as complete and 23 patients as incomplete. Two patients were not clearly defined as complete or incomplete. Fourteen patients had EC without intracardiac anomalies, 16 patients had intracardiac defects without EC, and 23 patients had both. Other associated anomalies were described in 29 patients. Thirty-seven of 58 patients, including patients in whom pregnancy was terminated, died within days after birth. This mortality from the reported literature may be higher because successful treatment of these patients is considered rare, and therefore these patients will be reported relatively more often. In this selected group, the mortality was higher in the patients with associated anomalies and if the complete form was present. The surviving patients with EC were those with associated intracardiac anomalies. This suggests that intracardiac defects may favor the prognosis. However, a selection bias was present due to the small number of patients.

In conclusion, the prognosis seems to be poorer in patients with the complete form of pentalogy of Cantrell, EC, and patients with associated anomalies. Intracardial defects do not seem to be a prognostic factor. When the diagnosis pentalogy of Cantrell is suspected, a multidisciplinary approach is essential. A prenatal medical team consisting of a gynecologist, a neonatologist, a pediatric cardiologist, a geneticist, and a pediatric surgeon should use their expertise in choosing the best approach to this severe disorder.

References

1. Abdallah HI, Marks LA, Balsara RK, Davis DA, Russo PA (1993) Staged repair of pentalogy of Cantrell with tetralogy of Fallot. Ann Thorac Surg 56:979–980
2. Alayunt A, Yagdi T, Alat I, Posacioglu H, Buket S (2001) Left ventricular diverticulum associated with Cantrell's syndrome and tetralogy of Fallot in an adult. Scand Cardiovasc J 35:55–57
3. Araujo Junior E, Zanforlin Filho SM, Guimaraes Filho HA, Pires CR, Nardozza LM, Moron AF (2006) Diagnosis of pentalogy of Cantrell by three-dimensional ultrasound in third trimester of pregnancy. A case report. Fetal Diagn Ther 21:544–547
4. Aslan A, Karaguzel G, Unal I, Aksoy N, Melikoglu M (2004) Two rare cases of the pentalogy of Cantrell or its variants. Acta Med Austriaca 31:85–87
5. Baker ME, Rosenberg ER, Trofatter KF, Imber MJ, Bowie JD (1984) The in utero findings in twin pentalogy of Cantrell. J Ultrasound Med 3:525–527
6. Bhat RY, Rao A, Muthuram (2006) Cantrell syndrome in one of a set of monozygotic twins. Singapore Med J 47:1087–1088
7. Bick D, Markowitz RI, Horwich A (1988) Trisomy 18 associated with ectopia cordis and occipital meningocele. Am J Med Genet 30:805–810
8. Bittmann S, Ulus H, Springer A (2004) Combined pentalogy of Cantrell with tetralogy of Fallot, gallbladder agenesis, and polysplenia: a case report. J Pediatr Surg 39:107–109
9. Bogers AJ, Hazebroek FW, Hess J (1993) Left and right ventricular diverticula, ventricular septal defect and ectopia cordis in a patient with Cantrell's syndrome. Eur J Cardiothorac Surg 7:334–335
10. Cantrell JR, Haller JA, Ravitch MM (1958) A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. Surg Gynecol Obstet 107:602–614
11. Carmi R, Boughman JA (1992) Pentalogy of Cantrell and associated midline anomalies: a possible ventral midline developmental field. Am J Med Genet 42:90–95
12. Chen CP, Hsu CY, Tzen CY, Chern SR, Wang W (2006) Prenatal diagnosis of pentalogy of Cantrell associated with hypoplasia of the right upper limb and ectrodactyly. Prenat Diagn 27:86–87
13. Chen Y, Lai HS, Duh YC, Wang JK, Chiu IS (1996) Pentalogy of Cantrell corrected by one-stage repair. J Formos Med Assoc 95:555–557
14. Correa-Rivas MS, Matos-Llovet I, Garcia-Fragoso L (2004) Pentalogy of Cantrell: a case report with pathologic findings. Pediatr Dev Pathol 7:649–652
15. Denath FM, Romano W, Solcz M, Donnelly D (1994) Ultrasonographic findings of exencephaly in pentalogy of Cantrell: case report and review of the literature. J Clin Ultrasound 22:351–354
16. Desselle C, Herve P, Toutain A, Lardy H, Sembely C, Perrotin F (2007) Pentalogy of Cantrell: sonographic assessment. J Clin Ultrasound 35:216–220
17. Egan JF, Petrikovsky BM, Vintzileos AM, Rodis JF, Campbell WM (1993) Combined pentalogy of Cantrell and sirenomelia: a case report with speculation about a common etiology. Am J Perinatol 10:327–329
18. Fox JE, Gloster ES, Mirchandani R (1988) Trisomy 18 with Cantrell pentalogy in a stillborn infant. Am J Med Genet 31:391–394
19. Grethel EJ, Hornberger LK, Farmer DL (2007) Prenatal and postnatal management of a patient with pentalogy of Cantrell and left ventricular aneurysm. A case report and literature review. Fetal Diagn Ther 22:269–273
20. Halbertsma FJ, van Oort A, van der Staak F (2002) Cardiac diverticulum and omphalocele: Cantrell's pentalogy or syndrome. Cardiol Young 12:71–74
21. Hornberger LK, Colan SD, Lock JE, Wessel DL, Mayer JE Jr (1996) Outcome of patients with ectopia cordis and significant intracardiac defects. Circulation 94:II32–II37
22. Hsieh YY, Lee CC, Chang CC, Tsai HD, Hsu TY, Tsai CH (1998) Prenatal sonographic diagnosis of Cantrell's pentalogy with cystic hygroma in the first trimester. J Clin Ultrasound 26:409–412
23. Knirsch W, Dodge-Khatami A, Bolz D, Valsangiacomo Buchel E (2006) Cantrell's Syndrome forme fruste in a newborn diagnosed by transthoracic echocardiography and cardiac magnetic resonance imaging. Pediatr Cardiol 27:652–654
24. Laloyaux P, Veyckemans F, Van Dyck M (1998) Anaesthetic management of a prematurely born infant with Cantrell's pentalogy. Paediatr Anaesth 8:163–166
25. Liang RI, Huang SE, Chang FM (1997) Prenatal diagnosis of ectopia cordis at 10 weeks of gestation using two-dimensional and three-dimensional ultrasonography. Ultrasound Obstet Gynecol 10:137–139
26. Marijon E, Hausse-Mocumbi AO, Ferreira B (2006) Cantrell's syndrome. Cardiol Young 16:95–96
27. Martin RA, Cunniff C, Erickson L, Jones KL (1992) Pentalogy of Cantrell and ectopia cordis, a familial developmental field complex. Am J Med Genet 42:839–841
28. McMahon CJ, Taylor MD, Cassady CI, Olutoye OO, Bezold LI (2007) Diagnosis of pentalogy of Cantrell in the fetus using magnetic resonance imaging and ultrasound. Pediatr Cardiol 28:172–175
29. Morales JM, Patel SG, Duff JA, Villareal RL, Simpson JW (2000) Ectopia cordis and other midline defects. Ann Thorac Surg 70:111–114
30. Nanda S, Agarwal U, Sen J, Sangwan K (2003) Cantrell's syndrome — report of two cases with one atypical variant. Arch Gynecol Obstet 268:331–332
31. Oka T, Shiraishi I, Iwasaki N, Itoi T, Hamaoka K (2003) Usefulness of helical CT angiography and MRI in the diagnosis and treatment of pentalogy of Cantrell. J Pediatr 142:84
32. Onderoglu L, Baykal C, Tulunay G, Talim B, Kale G (2003) Prenatal diagnosis of Cantrell's pentalogy: a case report. Turk J Pediatr 45:357–358
33. Pivnick EK, Kaufman RA, Velagaleti GV, Gunther WM, Abramovici D (1998) Infant with midline thoracoabdominal schisis and limb defects. Teratology 58:205–208
34. Polat I, Gul A, Aslan H, Cebeci A, Ozseker B, Caglar B, Ceylan Y (2005) Prenatal diagnosis of pentalogy of Cantrell in three cases, two with craniorachischisis. J Clin Ultrasound 33:308–311
35. Rashid RM, Muraskas JK (2007) Multiple vascular accidents: pentalogy of Cantrell in one twin with left sided colonic atresia in the second twin. J Perinat Med 35:162–163
36. Siles C, Boyd PA, Manning N, Tsang T, Chamberlain P (1996) Omphalocele and pericardial effusion: possible sonographic markers for the pentalogy of Cantrell or its variants. Obstet Gynecol 87:840–842
37. Song A, McLeary MS (2000) MR imaging of pentalogy of Cantrell variant with an intact diaphragm and pericardium. Pediatr Radiol 30:638–639

38. Soper SP, Roe LR, Hoyme HE, Clemmons JJ (1986) Trisomy 18 with ectopia cordis, omphalocele, and ventricular septal defect: case report. Pediatr Pathol 5:481–483
39. Spencer R, Robichaux WH, Superneau DW, Lucas VW Jr (2002) Unusual cardiac malformations in conjoined twins: thoracopagus twins with conjoined pentalogy of Cantrell and an omphalopagus twin with atretic ventricles. Pediatr Cardiol 23:631–638
40. Toyama WM (1972) Combined congenital defects of the anterior abdominal wall, sternum, diaphragm, pericardium, and heart: a case report and review of the syndrome. Pediatrics 50:778–792
41. Uygur D, Kis S, Sener E, Gunce S, Semerci N (2004) An infant with pentalogy of Cantrell and limb defects diagnosed prenatally. Clin Dysmorphol 13:57–58
42. Vazquez-Jimenez JF, Muehler EG, Daebritz S, Keutel J, Nishigaki K, Huegel W, Messmer BJ (1998) Cantrell's syndrome: a challenge to the surgeon. Ann Thorac Surg 65:1178–1185