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

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Abstract Two patients with incomplete pentalogy of Cantrell are described. The first was a girl with a large omphalocele with eversion 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 eversion 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. Intracardiac defects do not seem to be a prognostic factor.

Keywords Ectopia cordis · Pentalogy of Cantrell · Abdominal wall defect

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 eversion 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 charac-

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

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

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
| 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       | TOF, ASD                                            | None                                                                      | No       |
|                                | CF       | TOF, pulmonary atresia                             | None                                                                      | No       |
| Martin et al. 1992 [27]         | CF       | EC, hypoplastic left ventricle and atrium, dilated right ventricle, pulmonary valve and artery | 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       | VSD, double outlet right ventricle, bilateral superior vena cavae, pulmonary stenosis | None                                                                      | Yes      |
| Chen et al. 1996 [13]           | CF (?)   |                                                     | None                                                                      | Yes      |
| Homberger et al. 1996 [21]      | ?        |                                                     | 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       |
| Laloyaux et al. 1998 [24]       | CF       | VSD, ASD, tricuspid atresia, pulmonary stenosis     | Cystic hygroma                                                           | No       |
| 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       | VSD, LVD, ASD, TOF                                 | None                                                                      | Yes      |
| Alayunt et al. 2001 [2]         | CF (?)   |                                                     | None                                                                      | Yes      |
| Reference          | Form (conjoined twin) | 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; aortico pulmonary communication | Thoracopagus twins                                    | No       |
| Halbertsma et al. 2002 [20] | IF                  | LVD, ASD, VSD, anomalous venous pulmonary return                                | Omphalopagus twins                                     | No       |
| Nanda et al. 2003 [30] | CF                  | EC, VSD                                                                        | None                                                     | Yes      |
| Onderoglu et al. 2003 [32] | CF                  | EC                                                                             | None                                                     | 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, polypsplenia, segmentation defect of the lungs | No       |
| Uygur et al. 2004 [41] | IF                  | EC                                                                             | Left clubfoot, hypoplasticity 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       |
| Correa-Rivas et al. 2004 [14] | IF | EC                                                                             | Bilateral cleft lip and palate, bilateral pulmonary hypoplasia | No       |
| Polat et al. 2005 [34] | CF                  | EC                                                                             | Craniorachischisis, bilateral clubfoot                  | No       |
| Marjon et al. 2006 [26] | CF                  | EC                                                                             | Craniorachischisis, bilateral clubfoot and clubhand     | No       |
| Bhat et al. 2006 [6] | IF                  | LVD, VSD                                                                       | None                                                     | No       |
| Araujo Junior et al. 2006 [3] | CF | EC                                                                             | None                                                     | No       |
| Knirsch et al. 2006 [23] | IF                  | Mesocardia, VSD, ASD, LVD                                                        | None                                                     | Yes      |
| Chen et al. 2006 [12] | CF                  | EC                                                                             | Scoliosis, hypoplasia of the right upper limb, ectrodactyly of the right hand and foot | No       |
| Rashid et al. 2007 [35] | CF                  | EC                                                                             | Ecephalocele, 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 | Cardiac defect                                      | Associated anomalies                          | Survival |
|----------------------|------|----------------------------------------------------|-----------------------------------------------|----------|
| McMahon et al. 2007  | IF   | EC, TOF, VSD, hypoplastic pulmonary valve          | None                                          | ?        |
| Our patients         | IF   | EC, VSD                                           | None                                          | ?        |
|                      | 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 craniarachischisis [14, 29, 34]; limb defects such as clubfoot, absence of tibia or radius, and hypodactyly [33, 41]; and abdominal organ defects such as gallbladder 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. Intracardiac 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.

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