Crossed Pulmonary Arteries: Report of Two Cases with Emphasis on Three-dimensional Helical Computed Tomographic Imaging

Bang-Bin Chen,1 Hong-Jen Hsieh,2 Ing-Sh Chiu,3 Shyh-Jye Chen,1* Mei-Hwan Wu4

Crossed pulmonary arteries are due to an anomalous origin of both pulmonary arteries from the main pulmonary trunk. This anatomy is often associated with other congenital cardiac and extracardiac diseases. We report two neonates with complex congenital heart disease who had this disorder, which was detected during cardiac computed tomography (CT) with three-dimensional reconstruction but not during echocardiography or angiography. The first patient was a 3-day-old male neonate who had tachypnea and feeding problems since birth. Cardiac CT showed crossed pulmonary arteries, type B interruption of the aortic arch, a ventricular septal defect, and a large patent ductus arteriosus. He received an emergency T-colostomy at 3 days of age because of severe bowel distention. Low-type imperforated anus was diagnosed. His post-operative course was complicated with fluctuated saturation, seizure, hypocalcemia, hyperphosphatemia, and sepsis. Also found were cardiac defects, abnormal facies, thymic hypoplasia, cleft palate, hypocalcemia, and a variable deletion on chromosome 22q11 (CATCH 22 disorder). Because of his poor prognosis, the patient was transferred to another hospital on day 16 for further care, at the family's request. The other patient was a 5-day-old female neonate who had a heart murmur since birth. Cardiac CT showed crossed pulmonary arteries, truncus arteriosus, type A interruption of the aortic arch, a ventricular septal defect, an atrial septal defect, and a large patent ductus arteriosus. She received complete surgical correction, including division of the patent ductus arteriosus and repair of the other defects. Intermittent respiratory distress and decreased blood pressure complicated her postoperative course, and she died on the eighth day after surgery. Crossed pulmonary arteries complicated accurate interpretation of two-dimensional echocardiographs of the great vessels, as well as the course and location of catheters during cardiac catheterization. Three-dimensional CT provided a noninvasive approach to clearly recognize these malformations and the related anatomic structures. This information is important in planning and performing surgery in neonates with crossed pulmonary arteries. [J Formos Med Assoc 2008;107(3):265–269]

Key Words: computed tomography, crossed pulmonary arteries, pulmonary artery anomalies

Crossed pulmonary arteries are rare congenital anomalies characterized by an abnormal ostium of the left pulmonary artery that lies to the right of and above the right pulmonary artery. The branch pulmonary arteries then crisscross as they course to each respective lung. Recognition of this rare anomaly is important because it is often associated with other congenital heart disease and extracardiac anomalies. We report two neonates who had this anomaly along with complex congenital heart
disease, and we present what we believe are the first three-dimensional (3D) computed tomographic (CT) images of this condition.

Case Reports

Case 1
A cyanotic, tachypneic, full-term male neonate who weighed 2390 g at birth was referred to our hospital at 3 days of age after imperforate anus and congenital heart disease were diagnosed. The patient had tachypnea and feeding problems since birth.

Echocardiography demonstrated type B interruption of the aortic arch, a subaortic-malalignment ventricular septal defect (VSD), and a large patent ductus arteriosus (PDA) connecting from the pulmonary trunk to the descending aorta. Cardiac CT performed for further evaluation revealed a possible associated anomaly and showed results similar to the echocardiographic findings. However, CT demonstrated additional findings, such as crossed pulmonary arteries (Figure 1A), an aberrant right subclavian artery arising from the descending aorta, and absence of the thymus. A reconstructed 3D image clearly depicted the relationships between these abnormal vessels and their origins (Figure 1B).

The circulation was PDA dependent. Therefore, an intravenous infusion of prostaglandin was started. Because the patient had a severely distended abdomen and suspected perforation of the intestine, emergency T-colostomy was performed. The operation revealed only severely distended bowel loops with no evidence of perforation. Kidneys, ureters, and bladder imaging with urografin showed low-type imperforated anus.

The patient’s bowel sounds gradually improved, and his oral feeding was good 3 days after surgery. A pull-through procedure was scheduled for a later date. Fluctuated saturation of 60–90% was noted during his hospitalization, and his oxygen demand was difficult to taper. Seizures suddenly occurred on day 9 after the operation. The seizures subsided immediately after the patient was given lorazepam. His blood hemogram revealed leukocytosis with a leftward shift.

Figure 1. (A) Contrast-enhanced cardiac computed tomography shows interruption of the aortic arch and a large patent ductus arteriosus (PDA) connecting the pulmonary trunk (PT) with the descending aorta (DAo). The ostium of the left pulmonary artery (LPA) lies to the right lateral of and higher than that of the right pulmonary artery (RPA). (B) The surface shaded three-dimensional reconstructed computed tomography angiographic image from the right posterior view clearly depicts the origins and relationships between these abnormal great vessels. The aberrant right subclavian artery comes from the descending aorta. AAo = ascending aorta; ABSCA = aberrant right subclavian artery; IA = innominate artery; LAA = left atrial appendage; LCCA = left common carotid artery; RA = right atrium.
(white blood cell count, 26,250/µL; bands, 11%; segment, 80%), an elevated C-reactive protein level (3.26 mg/dL; normal range, <0.8 mg/dL), hypocalcemia (6 mg/dL; normal range, 2.1–4.7 mg/dL), and hyperphosphatemia (6 mg/dL; normal range, 2.1–4.7 mg/dL).

Lumbar puncture was performed to evaluate suspected meningitis, but the data were within normal limits. Calcium was administered, and the hypocalcemia improved gradually. Antibiotics were given under the impression of sepsis. Cardiac defects, abnormal facies, thymic hypoplasia, cleft palate, hypocalcemia, and a variable deletion on chromosome 22q11 (CATCH 22 disorder) were suspected, and results of a fluorescent in situ hybridization assay were positive for haploinsufficiency in chromosomal region 22q11. Because of his poor prognosis, the patient was transferred to another hospital on day 16 for further care, at the family’s request.

**Case 2**

A 5-day-old female neonate who weighed 3088 g at birth was evaluated for suspected heart disease because a grade III systolic murmur was heard over the left sternal border at birth.

Thoracic radiograph showed situs solitus, an enlarged heart, and increased pulmonary vascular markings. Two-dimensional echocardiography showed a type A interrupted aortic arch, a perimembranous VSD, a large PDA, a type II atrial septal defect, and mild aortic regurgitation. Cardiac catheterization showed the same findings, as well as severe obstruction of the left ventricular outlet. Cardiac CT showed additional findings, including crossed pulmonary arteries and truncus arteriosus. A reconstructed 3D image clearly demonstrated that the ascending aorta originated from the right lateral aspect of the truncus arteriosus and that the large PDA connected the pulmonary trunk to the descending aorta (Figure 2A).

Prostaglandin, dobutamine, furosemide, and digoxin were given. At the age of 8 days, the patient underwent complete surgical repair. She received division of the PDA and repair of the interrupted aortic arch, atrial septal defect, and VSD. Crossed pulmonary arteries were confirmed during surgery (Figure 2B).

![Figure 2.](image-url)
The day after the operation, the patient’s blood pressure decreased, and low oxygen saturation was noted. Surgery was performed to check for bleeding and revealed a minimal hematoma. Intermittent respiratory distress and decreased blood pressure complicated the patient’s postoperative course. Because of desaturation, she received cardiopulmonary resuscitation on postoperative day 7. However, the neonate died the next day.

Discussion

Crossed pulmonary arteries are the classic form of malposition of the branch pulmonary arteries, which Jue et al first described in 1966. In crossed pulmonary arteries, the ostium of the left pulmonary lies to the right of and superior to the right pulmonary artery. From their anomalous origins, the branch pulmonary arteries course to their respective lungs, crossing proximally. In most reported cases, malpositioning of the branch pulmonary arteries are diagnosed in association with truncus arteriosus and an interrupted aortic arch.

Both of our patients had type A or B interruption of the aortic arch and VSD. One patient also had truncus arteriosus. In addition, the first neonate had CATCH 22 disorder. The deletion in chromosomal region 22q11 may occur in patients with dysmorphic and cardiologic conditions, such as DiGeorge syndrome, velocardiofacial syndrome, and conotruncal anomaly face syndrome. CATCH 22 disorder is the most common interstitial deletion in humans, with an incidence of 1 case in 4000 live births. Numerous clinical findings have been reported in affected patients, including cardiac defects, characteristic facial features, thymic hypoplasia, cleft palate, hypoparathyroidism, learning difficulties, and psychiatric disorders. Therefore, comprehensive evaluation and follow-up are necessary for patients with 22q11 deletion.

The etiology of crossed pulmonary arteries has not been established. However, Jue et al proposed that this form of malposition might be due to counterclockwise rotation of the normally formed main and branch pulmonary arteries. This rotational abnormality may be due to faulty differential growth of the pulmonary trunk during or after complete septation of the truncus arteriosus. Recto et al described 10 patients in whom malposition of the branch pulmonary arteries was prospectively diagnosed with echocardiography; crossing was observed in four. DiGeorge syndrome was noted in five patients, and four had microscopic deletion of chromosomal region 22q11. This observation suggested that genetic evaluation should be considered in all patients with malposition of the branch pulmonary arteries. The authors thought that this malposition may involve a short main trunk of the pulmonary artery, which precludes pulmonary-artery banding as a palliative option because of the likelihood of iatrogenic stenosis of the right pulmonary artery.

Despite the abnormal location and course of the branch pulmonary arteries, crossed pulmonary arteries do not cause mechanical airway obstruction, and they are not associated with any hemodynamic abnormalities. In the literature, imaging modalities used to diagnose crossed pulmonary arteries include cardiac angiography, echocardiography, and magnetic resonance imaging. The unusual origin and course of the branch pulmonary arteries complicate accurate interpretation of both the location of the catheter and the relationship of the vessels during angiography. Suprasternal two-dimensional echocardiography has facilitated the detection of abnormalities of the great vessels, but diagnosis of this lesion is still difficult. Magnetic resonance imaging has the advantages of multiplanar views, but its cost and acquisition time limit its use in this setting.

Helical CT and its capacity for volumetric rendering enable noninvasive imaging of these complex anomalies. Transverse CT scans are particularly suited for depicting both the abnormal origin and the abnormal course of the left pulmonary artery. Owing to its orientation, the abnormal vessel can be completely hidden behind the proximal part of the right pulmonary artery on a frontal pulmonary angiogram; this obscuration accounts for some angiographic misinterpretations.
Helical CT angiography with 3D reconstruction is useful for identifying this anomaly in patients with complex congenital heart disease. In contrast to two-dimensional imaging, which requires special skills for interpretation, the unique 3D display from various angles may improve our understanding of patients’ complex anatomies. In addition, the short acquisition time and the requirement for minimal sedation make this technique practical to use in patients who are in an unstable condition. The 3D display is also useful to pediatric cardiac surgeons regarding preoperative simulation and postoperative assessment. High-resolution images can depict extracardiac structures without any limitations in the window due to the aortic arch, branch pulmonary artery, or PDA, for example. In our second patient, the crossed pulmonary arteries increased the difficulty of surgical repair of the adjacent great vessels, which might have affected the patient’s prognosis. Preoperative diagnosis of this disease can facilitate surgical recognition of this unusual anomaly. In our first patient, CT clearly outlined the pulmonary arteries and their relationship with the other great vessels and obviated cardiac catheterization.

This form of malposition of the branch pulmonary arteries should be distinguished from other conditions involving an anomalous origin of the branch pulmonary arteries, such as a pulmonary sling. In a pulmonary sling, the left pulmonary artery originates from the right pulmonary artery and courses between the esophagus and the trachea as it passes from the right hilum to the left lung. In summary, we report two neonates with crossed pulmonary arteries that were diagnosed during helical cardiac CT. The reconstructed 3D image was useful for visualizing this condition and for planning the surgical approach.

References