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TRUNCUS ARTERIOSUS IN A CASE OF SITUS INVERSUS WITH LEVOCARDIA

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Truncus arteriosus is a rare cardiac anomaly where the heart has a single outlet in the form of a common aorticopulmonary trunk. We report a rare association of truncus arteriosus with situs inversus and levocardia detected at autopsy in a full term infant who died 10 days after birth. Bahrain Med Bull 1996;18(2):

Truncus arteriosus is defined as a single arterial trunk leaving the base of the heart via a single arterial valve to supply directly the systemic, pulmonary and coronary circulations. It constitutes only 1 to 4 % of the cardiac malformations in large autopsy series[1]. The truncus arteriosus occurs due to a failure of development of the aorticopulmonary septum and is almost certainly linked to abnormal migration of cells into the heart from the neural crest[2]. Almost always there is a ventricular septal defect as a consequence of the absence of the infundibular septum. Most cases of truncus arteriosus die during the neonatal period or early infancy as a result of refractory congestive cardiac failure.

We report an extremely rare case, in that a truncus arteriosus was encountered in a neonate who also had situs inversus with levocardia.

THE CASE

A full term female infant weighing 2300 g was born by an uncomplicated vaginal delivery. The mother was a healthy 23 year old primigravida with no antenatal risk factors. The infant cried immediately after birth but had persistent central cyanosis with respiratory distress. Chest x-ray showed mild cardiomegaly. Ultrasonography showed situs inversus of the abdominal organs with levocardia. On echocardiography, atrial and ventricular septal defects with a right-to-left shunt were noticed. The pulmonary artery

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could not be visualised. Nine days after birth, the condition of the infant suddenly deteriorated with severe respiratory distress, and died on the tenth postnatal day. A complete autopsy was performed.

Autopsy findings: The infant was 45 cm in length, cyanosed, well built and well nourished. No external congenital anomalies seen. On examining the thoracic cavity, it was found that both the lungs were trilobed and well aerated. The heart was normal in position, but enlarged, weighing 20 g. Detailed dissection of the heart revealed a normal inferior vena cava. There was atrial situs solitus, i.e., the morphologically right atrium was situated to the right and the morphologically left atrium to the left. The foramen ovale was widely patent. The tricuspid and mitral valves were normal. The ventricular chambers were normal. A high ventricular septal defect was present. Both the ventricles were communicating with a common truncus arteriosus through this septal defect

(Fig 1). The truncal valve which was overriding the ventricular septum was tricuspid with normal cusps. The arch vessels were arising from the truncus. The posterior aspect of the truncus gave origin to a very narrow pulmonary artery which bifurcated close to the hilum of the lungs; one branch entering each lung. The coronary ostia and coronary arteries were normal.

Examination of the abdomen revealed complete situs inversus of the viscera which were morphologically normal (Fig 2). The brain was normal. Histopathologically, all the organs appeared normal.

A final diagnosis of truncus arteriosus with situs inversus and levocardia was made.

DISCUSSION

Joy et al[3] who studied 20 cases of isolated levocardia found arterial situs inversus in 15, indeterminate atrial situs in 2 and atrial situs solitus in only 3 cases. All these 3 cases had interruption of the inferior vena cava with azygous continuity. The associated intracardiac lesions were mild. Our case, in contrast, showed a normal inferior vena cava and severe intracardiac anomalies.

Levocardia in situs inversus is usually associated with "inversion" of the ventricles and abnormal connections of the great arteries[4]. In our case, both the ventricles were normally situated.

Collett and Edwards[5] classified truncus arteriosus into four groups:

Truncus type 1 - A single pulmonary trunk arises from the ascending common trunk and then bifurcates into right and left pulmonary arteries.

Truncus type 2 - The right and left pulmonary arteries have a separate origin from the back of the common trunk.

Truncus type 3 - Separate pulmonary arteries originate from the sides of the ascending trunk.

Truncus type 4 - The pulmonary blood supply originates from the descending aorta.

Our case was of Truncus type 1.

The truncal valve is most frequently tricuspid but may be bicuspid, quadricuspid, pentacuspid or even hexacuspid. It may show various abnormalities like stenosis, regurgitation and nodular thickening of the cusps due to an abundance of mucoid connective tissue[6]. Our case showed a tricuspid truncal valve which was morphologically normal.

Some cases of truncus arteriosus may show severe underdevelopment of the aortic arch manifested by interruption1. When there is interruption of the aortic arch, the descending aorta is supplied via a patent ductus arteriosus. Our case showed no arch abnormalities and the ductus was absent.

In our case the death of the neonate occurred due to congestive cardiac failure.

CONCLUSION

Truncus arteriosus is a rare cardiac anomaly and its incidence rate need to be estimated from large post mortem series.

REFERENCES

1. Freedom RM, Culham JAG, Moes CAF. Truncus arteriosus. In: Angiography of congenital heart disease. New York: The Macmillan Company, 1984:437-52.

2. William PL, Bannister LH, Berry MM, et al. Gray's anatomy. Edinburgh: Churchill Livingstone, 1995:1503.

3. Joy J, Kartha CC, Venkitachalam CG, et al. Isolated levocardia. An echocardiographic evaluation [Abstract]. Indian Heart J 1992;44:268.

4. Becker AE, Anderson RH. Dissection of the congenitally malformed heart. In: Pathology of congenital heart disease. London: Butterworths, 1981:29-43.

5. Collett RW, Edwards JE. Persistent truncus arteriosus. A classification according to anatomic types. Surg Clin North Am 1949;29:1245-70.

6. Becker AE, Becker MJ, Edwards JE. Pathology of the semilunar valves in persistent truncus arteriosus. J Thorac Cardiovasc Surg 1971;62:16-26.