Cor triatriatum dexter: unusual features in utero and after birth

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Abstract

Cor triatriatum dexter is a rare congenital anomaly, caused by the persistence of the right valve of the sinus venosus, which divides the right atrium into two chambers and its diagnosis is rather difficult, due to the features often mimicking Ebstein’s anomaly. We describe a case followed from the foetal age for a relative right ventricular hypoplasia and suspicious pulmonary stenosis and diagnosed after birth to have cor triatriatum dexter with an obstructing membrane in the right atrium, needing surgical correction, done successfully.

Keywords: Cor triatriatum dexter • Congenital heart disease • Foetal echocardiography

BACKGROUND

Cor triatriatum dexter is a rare congenital anomaly (less than 0.1% of all congenital heart anomalies), caused by the persistence of the right valve of the sinus venosus, which divides the right atrium into two chambers. Normally, this valve regresses early in the foetal life between 9 and 15 weeks’ gestation (w.g.). The membrane sometimes mimicks Ebstein’s anomaly [1, 2]. Frequently, the anomalies of the right-sided sections are associated. Some cases were diagnosed only in adulthood, often at operating table or at autopsy [3].

CASE REPORT

A 31-year old healthy primipara was found to have abnormal cardiac features during the routine obstetric scan at 22 w.g. Subsequently, mildly hypoplastic right ventricle and pulmonary stenosis were diagnosed at echocardiography in another centre. At 27 w.g., she was referred to our centre where we confirmed the presence of a moderately hypoplastic right ventricle, tripartite, with a normally inserted, non-dysplastic tricuspid valve presenting mild regurgitation (z-score of tricuspid annulus ~1.5) and a slightly abnormal pulmonary valve with systolic flow velocities at Doppler at upper limits for gestational age, consistent with the suspicion of a mild pulmonary stenosis. At 37 w.g., the dimensions of the right ventricle were unchanged, with a z-score of ~1.5 and the tricuspid valve presented only a tiny transvalvular flow and no regurgitation. The pulmonary valve was mildly thickened, with a systolic flow velocity at upper limits with a good size of annulus (3.5 mm—z-score ~2). Moreover, a large fluctuating membrane in the right atrium was seen moving close to the tricuspid valve; at this stage, this was considered to be the Eustachian valve or the Chiari’s network (see Fig. 1a and Supplementary Video 1 with a relative still frame).

A male infant was delivered at 37+ w.g., weighing 2850 g, Apgar 6/9, with the initial arterial saturation of 90% and bidirectional flow at foramen ovale and the duct. The right ventricle was moderately hypoplastic, tripartite, z-score ~1.2, with a reduced diastolic flow of the tricuspid valve and a mild protosystolic regurgitation. The pulmonary valve presented a smaller annulus (z-score ~2.6), without a significant gradient at Doppler. A redundant aneurysmatic membrane was seen fluctuating in the right atrium, reaching the foramen ovale and passing also through the tricuspid valve into the right ventricular cavity, with the flow at colour Doppler streaming both to the foramen ovale and through the tricuspid valve (see Fig. 1b and Supplementary Video 2 with a relative still frame). In differential diagnosis, we considered at first again the Eustachian valve and/or the Chiari’s network but finally we opted for the diagnosis of the cor triatriatum dexter in view of an apparent obstruction caused by the membrane. The pulmonary flow was unchanged, without a significant gradient and the duct remained open, with a bidirectional flow.

Prostaglandins were started after a few hours due to desaturation (80%).

A major desaturation (75–78%) occurred at 2–3 days, with a clear right-to-left shunting through the foramen ovale and the infant was sent for surgery. A large fibrous fenestrated membrane was seen in the right atrium arising close to the entry of the inferior vena cava; it was attached medially to the tricuspid valve annulus and to the lateral wall of atrium, with the caval flow conveyed to the foramen ovale. The membrane was excised and the foramen ovale sutured (Fig. 2). The saturation of the infant improved and the baby is now well at 9 months, with a normal saturation and with normalized dimensions of the right ventricle.

COMMENT AND CONCLUSIONS

In this case, our initial diagnosis was a variant of pulmonary stenosis with mildly hypoplastic right ventricle. However, the
association of hypoplastic right ventricle to only a mild–moderate pulmonary stenosis is rather unusual, as well as an abnormal tiny transtricuspid flow, instead of a dysplastic tricuspid valve and/or tricuspid regurgitation. A membrane within the right atrium seen in the third trimester of pregnancy was considered by us at first as the Eustachian valve or the Chiari’s network. Only after the birth, we were able to make a correct diagnosis, due to obstructive features of the membrane seen both at real-time echocardiography and by streaming of the flow at colour Doppler and in view of an increased desaturation of the baby.

In contrast, in two reports regarding paediatric cases, the cardiac features were initially considered as Ebstein’s anomaly with the Chiari’s network [1, 2], due to the displacement of the tricuspid valve by a moving membrane; their cases also had hypoplastic right ventricle, associated in the second one to the tricuspid regurgitation. On the contrary, in our case, the tricuspid valve was normally inserted, not dysplastic and with only a tiny flow into the right ventricular cavity. The membrane was apparently obstructing the right ventricular inflow and we can hypothesize that this fact very likely interfered with the development of the right ventricle. The normalization of the dimensions of the right ventricle after the surgical removal of the membrane seems to confirm this hypothesis.

The clinical manifestations of cor triatriatum are clearly dependent on the degree of septation of the right atrium and of obstruction. In the case of a relevant obstruction, post-natal desaturation appears, as in our infant and in one of the paediatric cases [1]. Also, another report describes a neonatal case presenting with cyanosis, in whom the initial suspicion was an anomalous pulmonary venous return [4]. With a minor degree of obstruction, the patients may remain clinically silent for some time as in the second paediatric case, recognized at 23 months [2] or even till the adulthood, as evident from some casual presentations. Two foetal cases associated with nuchal oedema were diagnosed at autopsy [5]. Different variants exist—sometimes associated with a true Ebstein’s anomaly and/or ostium secundum atrial septal defect [6–8], while one report regarding three neonatal cases describes a case associated with pulmonary atresia [9]. This variability contributes to the diagnostic difficulties. A correct
recognition of cor triatriatum dexter in symptomatic cases allows a surgical removal that resolves the clinical distress.

REFERENCES


