Topic 10 – Pediatric and congenital heart disease

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0228

Impact of the precision of prenatal diagnostic of congenital heart diseases on perinatal and long-term management

Myriam Bensemlali*, Meriem Mostefa Kara, Bettina Bessière, Fanny Bajolle, Jerôme le Bidois, Laurent Fermont, Damien Bonnet
APHP-Hôpital Necker-Enfants Malades, Paris, France
*Corresponding author: myriam.bensemlali@gmail.com (Myriam Bensemlali)

Objective To evaluate the impact of precising prenatal diagnosis of congenital heart diseases (CHD) on perinatal and long-term management.

Methods Over a 10-year period, 1258 neonates with a prenatally diagnosed CHD and 189 fetal autopsies after termination of pregnancy were included. Changes in CHD diagnosis were classified as totally different, similar but leading to changes in neonatal management, and similar without changes on initial management. The impact on long-term outcome was considered negative if the final diagnosis was a more complex CHD precluding the planned biventricular repair, or if additional surgical interventions were needed, or if the complexity level of the Aristotle score was increased. The impact on outcome was considered positive if biventricular repair was possible while not planned prenatally, or if the number of surgical interventions was reduced, or if the complexity level of the Aristotle score was lower.

Results The post-natal diagnosis was imprecise in 30.2% of the cases: completely different in 2.9%, led to changes in initial management in 8%, and did not affect initial management in 19.3%. Imprecision in the prenatal diagnosis had a negative impact on long-term outcome in 4.9% of the cases, and a positive impact in 4.1%.

In the fetal autopsy group (mean term 26 weeks), the diagnosis was imprecise in 54.5% of the cases: completely different in 8.5%, could have led to changes in postnatal management in 14.3%, and with minor differences that would not have led to changes in management in 31.7%. In both groups, the most frequent differences were anomalies of the outflow tract anatomy (43%), and the systemic or pulmonary veins (25%).

Conclusion Imprecision of prenatal diagnosis of CHD changes early management in 11% of the cases, and impacts long-term outcome in 9% of the cases. Improvement of CHD diagnosis for anatomy of the outflow tract and main veins should help to reduce impact on postnatal management and outcome.

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Left juxtaposition of the atrial appendages: where are the pectinate muscles?

Lucile Houyel* (1), Daniela Laux (2), Meriem Mostefa-Kara (2), Marie Gonzales (3), Bettina Bessières (3)
(1) Centre Chirurgical Marie Lannelongue, Le Plessis-Robinson, France – (2) APHP-Hôpital Necker-Enfants Malades, Paris, France – (3) APHP-Hôpital Armand-Trousseau, Paris, France
*Corresponding author: l.houyel@ccml.fr (Lucile Houyel)

Left juxtaposition of the atrial appendages (LJAA) is a rare anomaly in which the two atrial appendages (AA) are located to the left of the great arteries (GA). Although several anatomic studies have been published, the internal architecture of the right atrium (RA) in LJAA was not well described.

We reviewed 21 postnatal and 5 fetal human heart specimens with L-JAA. All had normal atrial situs, concordant atrioventricular and discordant ventriculoarterial connections: 20 D-transposition, 6 L-transposition. None had heterotaxy. There were 11 transpositions of the GA, 8 double outlet right ventricle, 3 double-inlet left ventricle, 4 tricuspid atresia. The conus was always abnormal, bilateral in 22, subaortic in 4. Pectinate muscles (PM) were confined inside the right AA (RAA) in 20/26 hearts (77%, group 1) and spilled out it without extending to the crux in 6 (group 2). In 9 of group 1 and 4 of group 2, there was a small accessory RAA in normal position with PM inside it (p=ns). The only significant difference between the 2 groups was the incidence of hypoplastic right ventricle (65% in group 1 vs 0 in group 2, p<0.03).

Conclusion In 77% of LJAA, PM do not extend to the crux and can be completely absent of the RA wall. This suggests that PM are constitutive of the morphologically RA only if the RAA is in normal position and raises several questions concerning the development of the atria. In other terms, the extent of the PM (to the crux of the heart or not) might be determined by the situation of the RAA relative to the great vessels, and thus would not be an intrinsic property of the morphologically RA. Normal atrial situs and absence of heterotaxy could indicate that outpouching of the AA from atrial walls might occur after the establishment of the left-right asymmetry, as assessed in one of the rare experimental models of LJAA. Further experimental studies are warranted to elucidate this anatomic and embryologic enigma.

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3D transthoracic echocardiography assessment of the pulmonary valve in patients with TOF

Khaled Hadeed*, Sébastien Hascoet, Romain Amadieu, Yves Dulac, Sophie Breinig, Philippe Acar
CHU Toulouse, Hôpital des enfants, Toulouse, France
*Corresponding author: hadeed.k@chu-toulouse.fr (Khaled Hadeed)

Background Accurate evaluation of pulmonary valve (PV) morphology and pulmonary annulus (PA) diameter is crucial before surgical correction of tetralogy of Fallot (TOF). Our aim was to assess PV morphology using three-dimensional transthoracic echocardiography (3D-TTE) in infants with TOF before surgical correction. And to compare PA diameter obtained by different imaging modalities.

Methods 30 patients with TOF were prospectively included. All patients underwent 2D and 3D-TTE, 23 patients underwent CT-Scan and 7 cardiac catheterization. PA diameter was measured using 2D-TTE in parasternal short axis view as recommended. 3D dataset was acquired using zoom mode at PV. Both vertical (Dv) and horizontal (Dh) diameters of PA were measured. Mean 3D diameter (3DD) was calculated as (Dv+Dh/2). Eccentricity index (EI) of PA was calculated (Dv—Dh/Dv). These measurements were compared to CT-Scan and angiography when available and to perioperative measurements.

Results Mean age 7.4 months (3-24 months), mean weight 6.6kg (4.5-13.5kg). PV was described as bicuspid in 15/30 patients by 3D-TTE from en face view, with 75% agreement between 3D-TTE and perioperative finding (20/30 patients). PA geometry was slightly asymmetric by 3D-TTE. Dv was significantly larger than Dh (8.4mm vs 7.4mm, p=0.001), and mean EI of PA was (10%). PA was more asymmetric in bicuspid valves rather than in tricuspid valves (EI 14% vs 7%, p=0.002). PA diameter didn't differ significantly between 2D-TTE, Dh, CT scan and angiography, and were significantly lower than mean 3DD and Dv. There was a very strong correlation between 3DD and perioperative measurement. PA was conserved in 53.3% without significant residual stenosis immediately and after a 20 month of follow-up (except 1 patient requiring balloon dilatation).

Conclusion 3D-TTE in patients with TOF is an accurate method to describe PV, and to measure PA size. Thus could help the surgical repair and the preservation of PV function.

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