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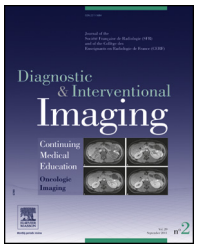

CONTINUING EDUCATION PROGRAM: FOCUS...

Imaging of congenital anomalies of the coronary arteries

Video Animation

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Abstract Congenital abnormalities of the coronary arteries are extremely variable and include anomalies of their origin (atresia, anomalies of origin from the aorta or from the pulmonary artery), the course of the epicardiac coronary branches (intramural, myocardial bridge) and distal connections (coronary-cardiac chamber fistulae). In pediatric practice, the diagnosis relies on ultrasound which should be supplemented by additional cardiac imaging in most cases. Multidetector CT is the most widely used imaging technique to identify abnormal courses and relationships with the greater vessels. In this paper, the important diagnostic and prognostic features in the interpretation of coronary imaging in pediatric practice is discussed.

Congenital anomalies of the coronary arteries are rare malformations, which must however be recognized firstly by pediatricians and, secondly, by adult cardiologists. Skills need to be acquired in order to identify pathological coronary artery appearances. This is also important in the training of radiologists who perform these types of investigations. Some congenital coronary anomalies have a typical presentation leading to a rapid diagnosis and decision on type of treatment. Others are suggested by atypical symptoms or are discovered incidentally during coronary imaging performed for other reasons.

In this article, we consider the isolated congenital malformations of the coronary arteries in which additional imaging is required after screening or a suspicious ultrasound. We then describe some types of acquired coronary artery anomalies in children after surgery for congenital heart disease with surgical reimplantation of coronary arteries.

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Embryological review

Initially, the vascular sinusoids develop within the embryonic myocardium before it becomes permanently compacted. At the same time, epicardiac blood islets enter, coalesce and form a rudimentary plexus. After a long period during which the origin of the coronary ostia was controversial, it is now known that they rejoin the aorta centripetally towards each of the corresponding sinuses of Valsalva. After the sinusoids have regressed and the epicardiac plexus has connected to the coronary sinus, the final arterial network is formed [1,2].

Definition of a congenital coronary artery anomaly

There is no specific definition other than the proposal by Abrams that a distribution or course should be considered as a variant rather than an anomaly if it is present in at least 1% of the population [3]. The presence of three coronary ostia (arising as a barrel from the left anterior descending and circumflex arteries) may be considered to be normal. Conversely, location close to the commissure of a right or left coronary ostium rather than a central position, even without clinical consequences, is liable to complicate, for example, an aortic valve replacement procedure. The initial course of the coronary arteries is usually perpendicular to the sinus, although a large range of orientations exist up to a tangential course which is considered to be abnormal. In addition, the distribution of the epicardiac arteries and their dividing branches is extremely varied and, again, defining normality is difficult.

Abnormalities of the origin of the coronary arteries

Ostial stenosis or atresia

Ostial stenoses are extremely rare anomalies of the coronary arteries with only a few clinical descriptions in the literature. Ostial atresia is seen particularly in association with cardiac malformations such as pulmonary atresia with intact septum, in which coronary perfusion may be supplied from the right ventricle through sinusoids. Its embryological origin is poorly understood but may be due to defective incorporation of the coronary bud into the corresponding sinus or defective formation of the proximal part of the vessel lumen. Angiographic diagnosis is difficult, particularly in young children with ischemic heart disease, and can only be achieved by demonstrating retrograde perfusion of the atretic vessel from the other coronary artery. The presence of a lavage, or jet, in the sinus of the Valsalva can distinguish stenoses from complete occlusions [4,5]. The differential diagnosis is with an ectopic coronary artery origin, particularly from the pulmonary artery which is considerably more common or extreme dominances which are the embryological equivalent of a single coronary artery [6]. The most common example of this type of lesion in children is, of course, postoperative acquired atresia following reimplantation of the coronary artery of abnormal origin or after an arterial switch for transposition of the greater vessels [7] (video 1).

Ectopic origin

This group contains a large range of coronary anomalies in which the origin of the artery concerned is not located in the middle of the corresponding sinus close to the sinotubular junction, but in an abnormal position arising from another sinus or from another coronary artery (single coronary artery) or another vessel (pulmonary arteries).

Abnormalities of origin from the aorta

Different types of abnormal origin from the aorta can be distinguished: commisural origin, origin from the ascending aorta above the sinotubular junction and origin from the contralateral sinus.

The prevalence of these abnormalities varies depending on whether the series are autopsies or angiographic. Typically, they are responsible for sudden death on effort or myocardial infarction, rarely preceded by angina. The assumed mechanism is compression of the inter-aorto-pulmonary tract during effort through dilatation of the cardiac great vessels or by progressive deterioration of the intramural course of the aberrant coronary artery towards stenosis [8].

Origin from the controlateral aortic sinus

The anomaly most typically described is a coronary artery arising from the controlateral sinus and following a course between the aorta and the pulmonary artery in the proximal epicardiac segment. This course may be located in the aortic wall and progress gradually towards main stem stenosis, which is not only functional on effort through distention of the root of the aorta but also organic as a result of internal remodeling in this abnormal portion [9–14].

Several types of aberrant courses are described: retro-aortic, inter-aorto-pulmonary, on the anterior wall of the right ventricle and infundibular or conal close to the pulmonary ring. Apart from the inter-aorto-pulmonary course which may be responsible for sudden death, these abnormal coronary branch courses are not of clinical significance.

The main types of path are described in Fig. 1.

In the majority of cases, this type of coronary anomaly can be diagnosed by echocardiography although a detailed analysis with coronary computed tomography (CT) or magnetic resonance imaging (MRI) is often required to specifically identify the anatomical features of the lesion which are required to define cardiac risk and treatment (Figs. 2–6). The risk is due to:

- the intervascular position of the aberrant coronary artery between the aorta and the pulmonary artery trunk (anterior loop, posterior loop, and conal septum courses are not considered to be at risk of an ischemic event);
- the type of coronary (the inter-aorto-pulmonary left coronary carries a higher risk of an ischemic event than an inter-pulmonary right coronary artery)
- the anatomical features of the proximal part of the aberrant coronary artery (narrow ''slit like'' ostium, acute angle of origin of the coronary artery from the aorta and aberrant intramural coronary course defined by an initial