Editorial Commentary

Coronary Anatomy in the Newborn:

What Do We Need to Know and When?

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These comments arise from issues raised by Robinson and colleagues[1] in their honest and diligent account (see pages 51-4) of their diagnostic evaluation of a stable and comfortable baby who was born with a heart murmur. In this case, the initial testing, performed at another hospital, included an echocardiogram that raised the question of a coronary anomaly, in addition to showing patent ductus arteriosus with a left-to-right shunt. To determine the validity of a suspected anomalous origin of the right coronary artery (RCA) from the pulmonary artery, the investigators performed 2 additional echocardiograms, and finally a left-sided heart catheterization. Once the diagnosis of ectopic origin of the RCA from the ascending aorta was made, the erroneous initial indication for urgent cardiac surgery was rendered null and void.

Although we appreciate the disciplined and educational presentation of the clinical case, we find lingering questions on the necessity, cost, and risk of these investigative procedures in a newborn baby, and on the nature and urgency of the various diagnoses at play.

Let us discuss the 2 central issues: 1) What types of coronary artery anomalies require surgery in a newborn? 2) What process is most reasonable in the management of a similar case of possible coronary anomaly in a newborn?

Critical Coronary Artery Anomalies in the Newborn. It must be agreed that the only coronary artery anomaly that definitely requires urgent surgery in the newborn or infant is anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA).[2,3] Extremely rare forms of congenital ostial atresia (COSA[4]) or stenosis of the left coronary artery are also possible, if less common, indications. In ALCAPA, it is typical that, at the time of maturation of the pulmonary arterial circulation, acute ischemia of the left ventricle manifests itself with acute electrocardiographic changes, cardiac failure, shock, and even death or severe cardiomyopathy, during the first month of life. Transthoracic echocardiography is usually an adequate diagnostic tool for this severe disorder. In the case under discussion, a tentative diagnosis of anomalous origin of the RCA from the pulmonary artery (ARCAPA)—a condition similar to, but much more benign than ALCAPA—could not be sustained by the clinical presentation, the electrocardiogram, or the echocardiogram. ARCAPA usually does not cause any of the clinical features of ALCAPA, nor does it generally constitute an indication for surgical correction in the newborn stage, when it is typically asymptomatic.

Most Reasonable Diagnostic Procedure in the Present Case. If ARCAPA was the important coronary artery anomaly to rule out, the initial left-to-right shunting (indicative of low pulmonary artery pressure) of the patent ductus arteriosus was by itself good negative evidence: ARCAPA could not have featured pro-grade flow (see Fig. 3B[1]) into the distal RCA. If the presence of pulmonary artery hypertension was a residual doubt after 3 echocardiograms (it does not appear to have been so), a quick right-sided heart catheterization would have been a sufficient and minimally interventional procedure, without deep general anesthesia. If that were the case, pulmonary hypertension would have been the dominant (and serious) issue.

The alternative possible but inconclusive diagnosis of anomalous origin of the RCA from the ascending aorta was apparently considered (in order to diagnose the cause of a murmur), in view of the later echocardiographic findings. Indeed, the detailed images (even though shown only as still images) indicate the presence of a large coronary ostium (Figs. 1A and 1B) at the ascending aorta, with pro-grade flow (Fig. 3B) suggestive of anomalous origin of the RCA from the ascending aorta. In addition, the

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proximal RCA appeared round and free of compression, in the absence of a tangential origin: indeed the RCA appears in Fig. 3A to diverge from the aortic wall, rather than follow an intramural course.

Even though this fact has not been widely reported in the literature,[2,3] ectopic origin of the RCA from the ascending aorta rarely features an intramural course with lateral compression (the condition for stenosis and ischemic manifestations[3]), which is observed in anomalous origin of the RCA from the left sinus of Valsalva. In addition, such ectopic origin has never been reported as a plausible cause of sudden cardiac death in a newborn—whereas anomalous RCA from the left sinus of Valsalva has indeed been reported (rarely) to cause sudden cardiac death in young men during sports activities.[2]

We should note, in particular, the following important reservations about performing arterial catheterization in a newborn in order to obtain coronary visualization: 1) substantial costs; 2) usually, the need for general anesthesia, and the attendant risks; 3) the definite risk of (at least) arterial-entry complications[5]; 4) the difficulty, in a newborn, of selectively catheterizing the ectopic RCA (which might clarify the ostial intramural course); and 5) the absence of a strict "need to know" the exact coronary anatomy of a newborn, in view of the above discussion.

Alternative imaging techniques for coronary visualization in the newborn could be coronary magnetic resonance or multidetector computed tomographic angiography (MDCT).[4,6] General anesthesia is usually indicated for both techniques, and the usual tachycardia in both instances would impede the detailed anatomic description of small and moving vessels. Moreover, MDCT requires significant doses of ionizing radiation.

In conclusion, the most prudent option in a case similar to that presented by Robinson and colleagues would probably be "watchful waiting," to be followed by exact anatomic description at a later time (adolescence), if clinically needed.

References

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