The black swan: Unique coronary arterial anatomy observed in a patient with transposition

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The unique coronary arterial anatomy found in a patient with transposition.

CASE REPORT

We describe a previously unreported arrangement of coronary arterial origin (Figure 1, A and B). The findings were encountered in a 22-year-old woman with transposition who had undergone a neonatal arterial switch procedure. She had been invited for routine postoperative in-hospital evaluation in accordance with our institutional protocol, which consists of computed tomographic angiography and magnetic resonance imaging.¹ She was asymptomatic, the results of all basic studies were within the normal range, and no evidence was found of ischemic changes. Tomography revealed a highly unusual coronary arterial pattern, which was significantly different than that described at the time of initial surgery (Figures 1, D, and 2). All 3 major coronary arteries arose separately from 1 sinus. The right coronary artery and the circumflex artery then coursed to the right side of the pulmonary trunk, with the anterior interventricular artery coursing to its left side. The right coronary artery then extended in anticipated fashion in to the right atrioventricular groove. The circumflex artery, however, looped anteriorly relative to the pulmonary trunk. It then crossed the anterior interventricular artery as it coursed toward the left atrioventricular groove. The anterior interventricular artery ran between the arterial roots. Its proximal interarterial course showed acute proximal angulation, with a high elliptical index, producing a moderate stenosis of 60%. In the initial surgical report, the circumflex artery had been confused with the anterior interventricular artery. Initially, the anterior interventricular artery had arisen from the right-handed aortic coronary sinus (sinus 1). Because of significant commissural malalignment (see Figure 1, B), both arterial buttons had been transplanted to the anterior adjacent sinus of the original pulmonary root. In the light of the high elliptical index, and evidence of 60% stenosis, we performed additional myocardial perfusion scintigraphy. This failed to reveal any significant resting or

Central Message

We describe a unique arrangement of the coronary arteries, which breaks the rule that the coronary arteries originate from the most convenient aortic valvar sinus.

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exercise-induced ischemia. Hence, we placed the patient in our group of survivors of the arterial switch procedure deemed to be at high risk, and requiring yearly clinical follow-up. Although at present consensus is lacking as to whether geometric findings revealed using computed tomography should dictate restriction of sporting activities, we provided additional recommendations regarding lifestyle and sporting activity according to our institutional protocol. We advocate that patients undertake mild-to-moderate physical activity on a daily basis, but we suggest restriction of severe or competitive sporting activity. We also recommend a healthy diet, adequate control of weight, and regular medical check-ups.

DISCUSSION

The various arrangements of the coronary arteries known to exist in the setting of transposition, when defined as the combination of concordant atrioventricular and discordant ventriculo-arterial connections, can all be described using the modified Leiden convention.² In most instances, the patterns are associated with given relationships of the transposed arterial roots. During the arterial switch procedure, the sinus origins of the arteries are transferred as button to the adjacent sinuses of the pulmonary trunk. The presence of unusual patterns, along with differences between

FIGURE 1. A, Three-dimensional volume rendered computed tomography reconstruction with separate color marking of each coronary artery. B, Schematic of actual postoperative anatomy determined using computed tomography and magnetic resonance imaging. C, Schematic of actual preoperative anatomy. D, Schematic of anatomy as it was reported by the surgeon at the time of the arterial switch operation. LA, Left atrium; MPA, major pulmonary artery; LV, left ventricle; LAD, left anterior descending artery (green); RA, right atrium; RCA, right coronary artery (violet); Cx, circumflex coronary artery (yellow); RV, right ventricle; AoV, aortic valve; PAV, pulmonary valve.

the levels of the roots, and commissural malalignment, can produce problems in proximal mobilization of the arterial stems. With some exceptions,³ most centers do not undertake routine postoperative coronary arterial imaging, nor is such a protocol supported in current guidelines.¹ On the basis of our experience, however, computed tomographic scans performed in young adults who have undergone a neonatal arterial switch procedure reveal abnormalities in half of otherwise asymptomatic patients. The clinical significance of these findings has yet to be established.¹ Findings of particular potential concern are acute proximal angulation, a high elliptical index, and an interarterial course,¹ however, this selection was mainly on the basis of studies related to patients with isolated coronary anomalies who are otherwise healthy. Such features might lead to coronary stenosis and future ischemic complications. Patients with such findings require watchful follow-up, with restriction of some competitive sporting activities. The

identification of such patients can be made only on the basis of direct imaging.

The significance of the unique arrangement encountered in our patient is that it breaks the hypothetical rule related to the epicardial course of the coronary arteries and their sinusal origin.⁴ The rule is on the basis of the notion, still maintained in the modified Leiden convention, that one coronary artery will not cross the territory of another artery.² The theory was developed on the basis of the known existing patterns for sinusal origin. The best way of disproving any hypothesis, such as “all swans are white,” is to provide evidence of the black swan. Our patient qualifies as the “black swan.” In most patients with the epicardial course as observed in our patient, the circumflex artery, having taken its origin from sinus 2, courses in the transverse sinus posterior to the pulmonary trunk to reach the left atrioventricular groove. This pattern is the second most frequent arrangement found in the setting of transposition. The

FIGURE 2. Two-dimensional curved multiplanar reconstruction of the coronary arterial anatomy with particular focus on the sinusal origins of the main epicardial coronary arteries: right (RCA) and circumflex (Cx) coronary arteries (white circle), left anterior descending (LAD) artery (blue circle), and interarterial pattern of LAD with height-to-width ratio (green circle). MPA, Major pulmonary artery; Ao, aorta; LA, left atrium.

crossing of territories in our patient breaks this notion of direct origin between a given epicardial coronary artery and its sinusal origin. The finding also provides evidence of the existence of a pattern previously considered to be

“impossible” on the basis of the existing hypothesis.⁴ Recent findings regarding the development of the coronary arteries, nonetheless, do provide a rational explanation for the unexpected finding. Rather than growing into the arterial sinuses, as has previously been presumed to be the case, the recent evidence shows that the coronary arterial stems grow out of the developing sinuses.⁵ This means that a stem could grow in an unexpected fashion, crossing over an additional stem growing from the same sinus, with the crossing stems then uniting with the major coronary arteries developing separately within the epicardium.

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