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IMAGING VIGNETTE

A CT Study of Coronary Arteries in Adult Mustard Patients

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COMMON TRANSPOSITION OF THE GREAT ARTERIES (TGA), defined as the aorta arising from the morphological right ventricle and the pulmonary artery arising from the morphological left ventricle, is a lethal and relatively frequent malformation, accounting for 5% and 7% of all congenital heart disease. The atrial switch operation, known as a Senning or Mustard procedure, was the surgical treatment for TGA used until the late 1980s. This procedure creates a physiological correction of this defect by baffling the systemic veins to the left ventricle and of the pulmonary veins to right ventricle. The atrial switch dramatically improved life expectancy for TGA and the patients who benefited are now reaching adulthood. Many studies have been published about long-term follow-up in Mustard/Senning patients with dysfunction of systemic right ventricle as the main problem. The pathophysiology of right ventricular failure is not fully understood. Some studies have suggested an ischemic pathogenesis, but we are aware of no report of systematic evaluation of coronary artery anatomy in adult Senning or Mustard patients (1).

In these adult patients coronary angiography imaging can be complicated by the variability and complexity of cardiac anatomy, modification of anatomy by surgery, and interference from devices such as conduits, baffles, stents, and coils. Coronary computed tomography angiography (CTA) is a new and rapidly emerging noninvasive imaging technique for the study of the coronary arteries in adult patients, with results equal, if not superior, to coronary angiography for depicting coronary artery anatomy. Radiation dose remains the only limitation to the use of this technique.

We performed coronary CTA in a group of 26 Mustard patients (mean age 22 ± 3 years; range 14 to 26 years) using a 64-slice scanner (Aquilon 64, Toshiba Medical Systems, Nasu, Japan) with a retrospective electrocardiography-gated data acquisition protocol. All patients had a pacemaker so that their heart rate could be controlled by the device—which was set to a heart rate of 75 beats/min—without administration of beta-blocking agents before the computed tomography scan. In most cases coronary CTA scans were obtained from the carina to the diaphragmatic surface of the heart using an automatic starting protocol ("sure start") with the region of interest positioned at the level of the right ventricle and triggered at 150 Hounsfield units. Ninety ccs of nonionic, iodinated contrast media was injected via the antecubital vein through an 18-gauge needle followed by 60 ccs of normal saline, both at 4.5 cc/s. The radiation dose for exam was calculated to be in the range of 8 to 10 mSv.

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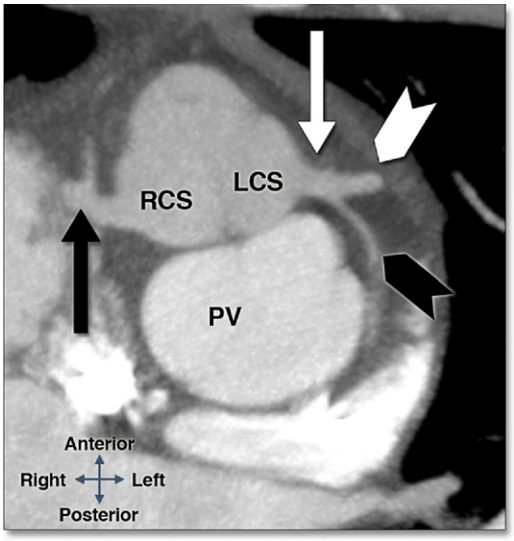


Figure 1. Usual Coronary Pattern for TGA

An oblique 3 mm maximum intensity projection (MIP) image of the usual coronary pattern for transposition of the great arteries (TGA), observed in most of our patients (12 of 26 patients). The left main stem (LMS) **(white arrow)** arises from the left coronary sinus (LCS) (the sinus adjacent to the pulmonary valve [PV] more leftward) and divides into left anterior descending coronary artery (LAD) **(white arrowhead)** and a left circumflex coronary artery (LCX) **(black arrowhead)**. The LAD passes anterior to the PV to reach the anterior interventricular groove and the LCX passes anterior and lateral to the PV to reach the posterior left atrioventricular groove. The right coronary artery (RCA) **(black arrow)** arises from the right coronary sinus (RCS) (the sinus adjacent to the PV more rightward) and passes anterior to the tricuspid valve to reach the right atrioventricular groove. In this case the aorta is anterior to the PV as observed in most of our patients with the usual coronary pattern. None of our patients with the usual coronary pattern had the aorta posterior to or side by side with the pulmonary artery.

The results of these CTA studies are shown in Figures 1, 2, 3, 4, 5, and 6 (2) and they include considerable variability in the anatomy of coronary arteries that could be important for planning diagnostic or therapeutic procedures and show hypoplasia of the left coronary artery system. We described the length and size of coronaries according to the following criteria: right coronary artery as normal when it reached and passed into the anterior atrioventricular groove otherwise hypoplastic; left main stem as normal or very short/absent; left anterior descending coronary artery as normal when it reached at least the mid-portion of the interventricular groove otherwise hypoplastic; left circumflex coronary artery (LCX) as normal when it reached and passed into the posterior atrioventricular groove otherwise

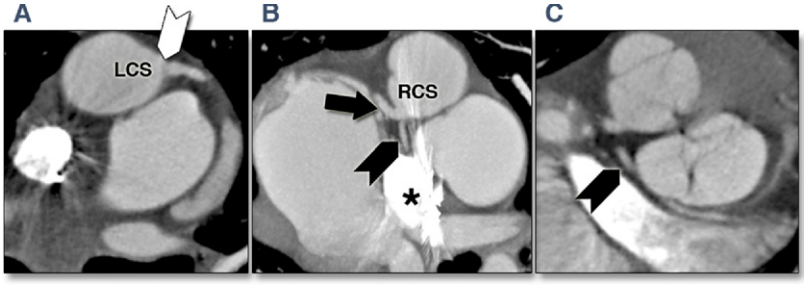


Figure 2. A Common Variant Coronary Pattern for TGA: Inverted Origin of LCX

Oblique MIP images of a patient with inverted origin of LCX, the most frequent unusual pattern observed in our patients (5 of 26 patients). In **panel A**, only the LAD **(white arrowhead)** is recognized arising from the LCS, passing anterior to the PV to reach the interventricular groove. The coronary artery arising from RCS bifurcates immediately **(B)** into the RCA **(black arrow)** and the LCX **(black arrowhead)**. The course of the LCX behind the PV is better seen in **(C)** where the artefact from the pacemaker (*) is less prominent. Abbreviations as in Figure 1.

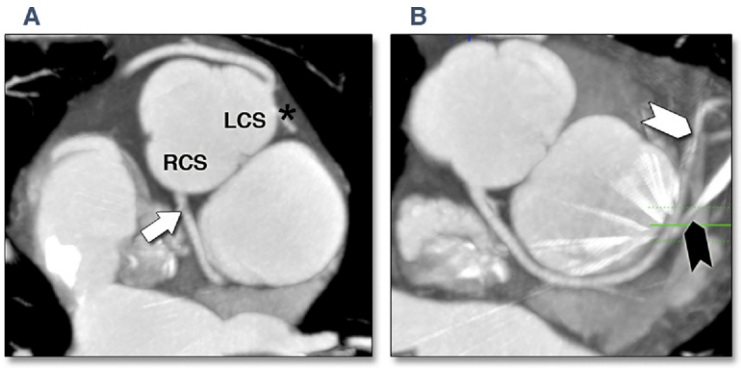


Figure 3. An Unusual Coronary Pattern for TGA: Totally Inverted Origin of Coronaries

An oblique MIP reconstruction of a case of inverted coronary arteries, a pattern observed in 2 of 26 patients. In **panel A**, RCA arises from the usual LCS and gives off a small conal branch (✻) before passing anterior to the aorta heading to the right atrioventricular groove. In the same image, the left coronary artery (LCA) arises from the usual RCS and passes posterior to the PV to reach the left atrioventricular groove **(white arrow)**. In **panel B**, the more distal course of the LCA and the bifurcation into the LCX **(black arrowhead)** and LAD **(white arrowhead)** are better defined. Abbreviations as in Figure 1.

hypoplastic. Hypoplasia was also defined by sizing the diameter of the coronaries with a value of >4 mm for men and >3 mm for women as normal range (3). The results of this classification are shown in Table 1.

A right dominant pattern, an absent or very short LMS and a severe hypoplasia of LCX in the majority of our patients (diameter medium of LCX 0.9 mm, range 0.5 to 2.0 mm) were the most peculiar findings.

Conclusions

64-slice coronary CTA is an excellent noninvasive technique for detailed study of coronary anatomy in adult Mustard patients. The epicardial course of the coronary arteries should be defined prior to interventional procedures. The left coronary system was hypoplastic in the majority of our patients and might cause ischemia of the left ventricle, with important implications for late retraining or biventricular resynchronization. Further studies are necessary to confirm this interesting hypothesis.

Table 1. Hypoplasia of Left Coronary System

	Size of Coronaries	
	Normal	**Hypoplastic/Absent**
RCA	26 of 26 patients (100%)	—
LMS	9 of 26 patients (35%)	17 of 26 patients (65%)
LAD	23 of 26 patients (88%)	3 of 26 patients (12%)
LCX	9 of 26 patients (35%)	17 of 26 patients (65%)

LAD = left anterior descending coronary artery; LCX = left circumflex coronary artery; LMS = left main stem; RCA = right coronary artery.

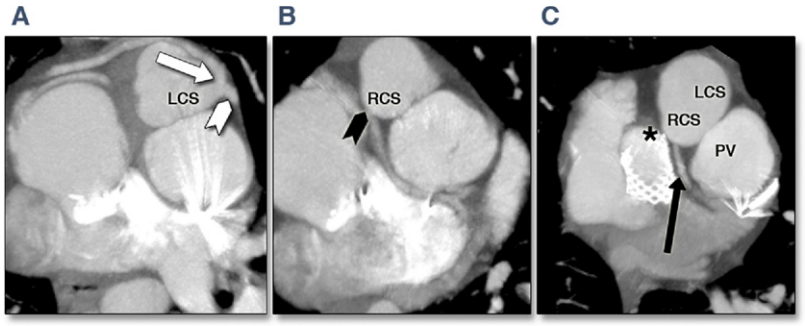


Figure 4. A Rare Coronary Pattern for TGA: Inverted RCA and LCX

An oblique MIP reconstruction of a patient with inversion of the RCA and the LCX, a pattern observed in 2 of 26 patients. The RCA **(white arrow)** and LAD **(white arrowhead)** arise together from the usual LCS **(A)**. The RCA then passes anterior to the aorta to anterior to reach the right atrioventricular groove while the LAD follows its usual course to the anterior interventricular groove. The LCX **(black arrowhead)** arises from the usual RCS and passes behind the pulmonary valve into the left atrioventricular groove **(B)**. **Panel C** is an oblique MIP reconstruction of the same patient at level of the superior vena cava showing the anomalous LCX **(black arrow)** following a retropulmonary course, close to a stent **(*)** in the superior limb of the systemic venous baffle. Although the LCX was not compressed in our case, it points out the importance of knowing the proximal courses of the coronary arteries in patients with TGA prior to undertaking interventional procedures at the base of the heart. Abbreviations as in Figure 1.

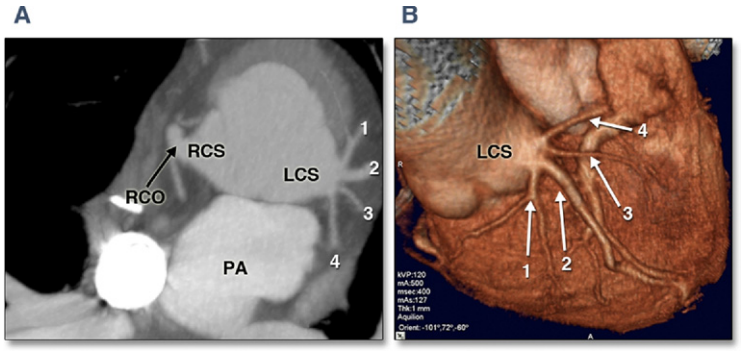
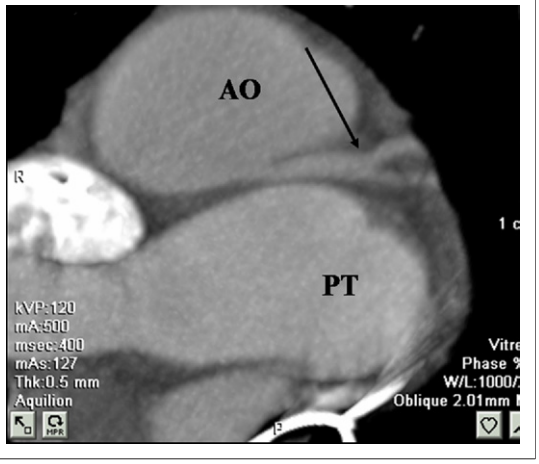


Figure 5. A Case of Multiple Ostia

Coronary computed tomography angiography in our patients has detected that multiple orifices and anomalies, such as absent or short LMS, can make coronary cannulation difficult and can increase the risk of coronary dissection or failure to completely delineate the coronary anatomy. This patient has multiple ostia of the left coronary system. MIP **(A)** and 3D volume rendering **(B)** reconstructions show a very peculiar aspect of the left coronary system with a right ventricular outflow branch **(1)**, LAD **(2)**, intermedius branch **(3)**, and LCX **(4)** arising separately from the LCS. PA = pulmonary artery; RCO = right coronary ostium; other abbreviations as in Figure 1.

Figure 6. Interarterial Course of Coronary Vessels

An oblique MIP image shows a potential course of LMS **(black arrow)** between aorta (AO) and pulmonary trunk (PT). The vessel originates at an acute angle with the wall of the aorta. These anatomic details could cause exercise induced ischemia because this segment of coronary could be squeezed between the pulmonary artery and the aorta as they dilate due to increasing blood pressure and flow during effort (2). Abbreviations as in Figure 1.



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