# Case report: echocardiographic detection of rare single coronary artery anomaly in a child with an atrial septal defect

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## Background
A single right coronary artery (RCA) with the left anterior descending (LAD) and circumflex coronary arteries located in the usual anatomic position and supplied by collaterals is the rarest variant of single RCA.

## Case summary
We report a paediatric patient with an incidental finding of single RCA Lipton type RI pattern during assessment for transcatheter device closure of an ostium secundum atrial septal defect (secASD). Transthoracic echocardiography (TTE) revealed a dilated RCA, abnormal flow in the LAD, and no identifiable left main coronary artery. Diagnosis of a single RCA was confirmed with angiography. Dobutamine stress echocardiography revealed no inducible ischaemia. Transcatheter device closure of the secASD was subsequently successfully performed.

## Discussion
TTE in paediatric patients can raise suspicion of coronary artery origin anomalies. Additional modalities, such as computed tomography and angiography, are required to comprehensively determine coronary artery anatomy. Functional assessment of ventricular function is also indicated. Coronary artery anatomy is important to delineate prior to transcatheter device closure of a secASD and should be part of the pre-procedure assessment.

## Keywords
- Single right coronary artery
- Atrial septal defect
- Coronary artery anomaly
- Case report

## Learning points
- Detecting coronary anomalies and defining coronary distribution is necessary prior to transcatheter secundum atrial septal defect closure.
- A single right coronary artery (RCA) with the left anterior descending and circumflex in the usual anatomic position being supplied by collaterals is the rarest variant of single RCA, also known as Lipton type RI pattern.
- Suspicion of coronary anomalies can be raised by transthoracic echocardiography in paediatric patients; cross-sectional imaging is required to delineate comprehensive coronary anatomy.

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## Introduction
A single right coronary artery (RCA) with left anterior descending (LAD) and left circumflex (LCX) coronary arteries located in the usual anatomic position and supplied by collateral arteries, also known as type RI using the Lipton et al.\(^1\) classification is the rarest congenital single coronary artery variant. A large study of 215 140 patients undergoing angiography found an incidence rate of 0.002%.\(^2\)

Transthoracic echocardiography (TTE) in paediatric patients is superior for coronary origin imaging resolution and assessment compared to adult patients. Coronary origin anomalies are important to identify...
as some variants can have clinical consequences. This case reports on the incidental TTE finding of the Lipton type R1 coronary artery anomaly in a child referred for an ostium secundum atrial septal defect (secASD) closure.

Timeline

| Time       | Event                                                                 |
|------------|----------------------------------------------------------------------|
| 5 months prior | Patient diagnosed with ASD at a visiting cardiology clinic and referred to paediatric cardiac centre overseas |
| Day 1      | Transthoracic echocardiogram confirming atrial septal defect suitable for transcatheter device closure  |
|            | Suspicion of single right coronary artery (RCA) anatomy raised         |
| Day 2      | Pre-catheter transoesophageal echocardiogram confirming secundum atrial septal defect suitable for transcatheter device closure, no left main coronary origin seen. Aortic root angiogram and subsequent selective right coronary angiogram demonstrating Lipton type R1 single RCA anatomy Dobutamine stress echocardiogram confirming no inducible ischaemia |
| Day 15     | Successful transcatheter atrial septal defect closure undertaken     |

Case presentation

A 9-year-old male patient with severe intellectual impairment and limited mobility was referred to local cardiology services with a soft ejection systolic murmur. The electrocardiogram showed an incomplete right bundle branch block with QRS duration of 95 ms and right axis deviation. Chest X-ray results showed mild cardiomegaly. The patient was diagnosed with a large secASD with evidence of right ventricular volume overload based on a focused TTE. Foetal alcohol syndrome was determined as the most likely overriding aetiology for his cardiac abnormality and co-morbidities. The patient resided in the South Pacific islands and was referred to the regional overseas paediatric cardiac centre for transcatheter secASD closure.

A full cardiac anatomy scan was completed using TTE at the cardiac centre. A diagnosis of secASD suitable for transcatheter device closure was confirmed. The patient had normal left ventricular dimensions and ejection fraction with no evidence of regional wall motion abnormalities. The right ventricle was severely dilated and had normal systolic function. Trivial mitral regurgitation was present. The patient had a severely dilated RCA origin from the right aortic sinus that measured 4.6 mm with a Boston Z-score of +4.65 (Figure 1A and Video 1). There was no evidence of a left main coronary artery (LMCA) originating from the left aortic sinus (Figure 1B and Video 2). On further interrogation using colour Doppler, only retrograde flow was noted in the region of the proximal LAD (Figure 1C and Video 3). The LCX was not observed.

The coronary artery origins were further assessed using transoesophageal echocardiography (TOE) due to suspicion of a coronary artery abnormality. This was undertaken as part of the secASD imaging pre-intervention. An aortic angiogram was chosen as the next step if a coronary artery anomaly could not be ruled out. The TOE was unable to identify an LMCA and demonstrated abnormal coronary artery colour Doppler signal near the left atrial appendage (Figure 1D and Supplementary material online, Video S1).

An aortic root angiogram showed a single dilated RCA originating from the right aortic sinus, supplying the inferoseptal and inferior myocardium and retrogradely filling the LCX back to the expected position of the LMCA. There was no LMCA originating from the aorta (Figure 2A and Supplementary material online, Video S2).

A selective RCA angiogram was subsequently performed to further define coronary supply and confirmed the above findings (Figure 2B and Supplementary material online, Video S3). There was adequate retrograde filling of the LCX as far as the LAD and LCX bifurcation. No LMCA was evident. The LAD was antegrade filled from the LCX and retrogradely by a large collateral artery from an RCA ventricular branch that extended to the cardiac apex. The middle third of the LAD was diffusely hypoplastic.

A dobutamine stress echocardiogram (DSE) was immediately performed to determine the potential need for surgical coronary artery intervention. DSE analysis confirmed good left ventricular function with no evidence of regional wall motion abnormalities.

The transcatheter secASD closure was deferred pending cardiovascular evaluation of the coronary artery findings. The overall appearance was judged to be a congenital coronary artery anomaly, but the diffusely hypoplastic LAD raised the question of inflammatory, and thus acquired aetiology.

It was decided that the risks of surgical coronary artery intervention to bypass graft or re-anastomose the left coronary arteries to the aortic root outweighed the benefits. Contributing factors to this decision included no inducible ischaemia or wall motion abnormalities, adequate collateral and retrograde supply, distance from the aortic root to the reconstitution of the LAD and LCX, and the small calibre of both the LAD and obtuse marginal branch. An interval transcatheter secASD closure was successfully performed using a 24-mm Figulla Flex II ASD occluder (Occlutech GmbH, Jena, Germany), and the patient then returned to his country of origin. A follow-up stress echocardiogram will be performed in adolescence.

Discussion

While single coronary artery anomalies are rare, they may have prognostic significance3,4 and are commonly associated with additional cardiac defects.4 The single coronary artery classification system by Lipton et al.1 denotes the ostium location (R: right coronary sinus, L: left coronary sinus) and uses a grouping system based on the anatomical distribution (I: RCA, LAD, and LCX are in the usual location connected by collateral circulation; II: proximal segment of single coronary artery gives rise to coronary artery trunk that supplies the
contralateral coronary artery; and III: LAD and LCX arise separately from proximal single RCA). Group II further categorizes the anatomy by the course of the transverse trunk in relation to the great arteries (A: anterior, B: between, P: posterior). A single RCA variant with interarterial course (Lipton RII-B) is the most serious, with a potential for sudden cardiac death. The present patient had the rarest single RCA variant of Lipton type RI, which has an incidence rate of 0.002%.

A genetics review determined that the patient fulfilled some but not all of the phenotypic criteria for both Prader–Willi and Cohen syndromes. There was sufficient evidence to support Foetal alcohol syndrome, making it the most likely aetiology. Foetal alcohol syndrome has a strong association with congenital heart defects. A review of 56 patients with Foetal alcohol syndrome found that 16 patients had evidence of congenital heart disease. The most common type of congenital heart disease in this group was an atrial septal defect.

Figure 1 Transthoracic and transoesophageal echocardiography of single coronary artery. (A) Two-dimensional transthoracic echocardiography image in parasternal short-axis view shows a dilated right coronary artery measuring 4.6 mm with a Boston Z-score of +4.65. (B) Transthoracic echocardiography colour comparison image of parasternal short-axis with focus on left aortic sinus with no left main coronary artery identified. (C) Transthoracic echocardiography colour comparison of left anterior descending region with abnormal flow. (D) Transoesophageal echocardiography image of retrograde flow in left circumflex artery towards junction with left anterior descending in the region of left atrial appendage.

Video 1 Transthoracic echocardiography of the aortic root en face showing a severely dilated right coronary artery.
Coronary artery anatomy is important to identify prior to transcatheter secASD closure. As an example, a retro-aortic circumflex artery off the RCA can be compressed by a secASD device, leading to ischaemia. In the present case, no LMCA had been identified on TTE or TOE images. It was important to confirm that coronary artery anatomy would be suitable for secASD device placement. Thus, coronary artery angiography was performed. In addition, a coronary artery anomaly necessitating surgical intervention would support a concomitant surgical closure of the secASD, rather than a separate device closure. Angiography results demonstrated mild hypoplasia of the mid-LAD. Due to the absence of regional wall motion abnormalities on resting TTE, inducible myocardial ischaemia needed to be excluded. A DSE was performed under the same anaesthetic because the patient was unable to manage exercise stress testing. According to the American Society of Echocardiography Stress Echocardiography Guidelines, the use of dobutamine stress echocardiography to detect inducible ischaemia in paediatric patients has increased. It has advantages in paediatric patients with both congenital and acquired coronary artery anomalies, because it avoids radiation exposure and provides a direct comparison with the resting ventricular function. DSE has an advantage over exercise stress testing, as it maintains an increased heart rate for a longer time, especially in children.

Although coronary artery anomalies are typically diagnosed via coronary artery angiography, computerized tomography, or at autopsy, TTE is capable of detecting coronary artery abnormalities in children. It should be routine to identify paediatric patients’ coronary artery origins as part of a full echocardiographic study. Coronary artery diameter can be normalized to body surface area in paediatric patients when the artery appears dilated. To optimize assessment of coronary arteries by TTE, the patient needs adequate acoustic windows and the echocardiographer needs to be experienced in coronary artery imaging. A transducer with the highest possible frequency and optimized settings should be used. Two-dimensional (2D) gains should be lowered and the zoom feature utilized or sector width decreased to optimize the frame rate. The colour Doppler scale should be set to between 20 and 40 cm/s and the colour box kept narrow to achieve the highest possible colour frame rate. Switching to a lower-frequency transducer may be necessary to demonstrate adequate colour Doppler signal for coronary
artery filling. Left parasternal imaging one to two rib spaces higher than standard can be helpful to enhance 2D TTE left coronary artery views.

**Lead author biography**

Rachel Hazaert is a 36-year-old cardiac sonographer who has worked in paediatric cardiology for 8 years. She trained and worked at Boston Children’s Hospital. She currently works at Starship Children’s Hospital, in Auckland New Zealand. She performs paediatric and congenital transthoracic echocardiography. She performs a cardio-surgical audit, presenting findings to the department biannually.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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