Twelve-month-old girl with myocardial ischaemia

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A 12-month-old girl was admitted to Kanagawa Children’s Medical Center because of the diagnosis of anomalous origin of the right coronary artery from the main pulmonary artery (ARCAPA). The electrocardiogram (ECG) recorded non-significant findings at rest but the ST depression from V1 to V3 at her crying.

On the coronary angiography, it looked like a single coronary artery from the left coronary sinus; however, this artery was dilated and meandering and linked the RCA whose flow was retro-perfusion, that is, continuous flow poured into proximal of the main pulmonary trunk beside the aortic root, namely, the original site of RCA (Figure). The operation confirmed the budding and meandering conus branch of the RCA on the right ventricular outflow tract and linked the RCA originated from the main pulmonary trunk to the left anterior descending artery. Re-implantation was performed using cardiopulmonary bypass, and the anomalous artery was anastomosed with the ascending aorta that was hinged with a J-shaped incision. The postoperative course was excellent. When last seen, 20 months after the operation, she was well with no evidence of myocardial ischaemia.

ARCAPA is rare. To the best of our knowledge, it had been reported that there were only three cases of isolated ARCAPA reported under 2 years of age. The reason is many of these cases were reported not lethal or asymptomatic, in contrast to the origin of the left coronary artery from the pulmonary artery. However, it has some reported sudden death ARCAPA cases revealed by autopsy. In our case, ECG demonstrated myocardial ischaemia at the time of her crying.

Figure. Angiography: pre-operative. left coronary arteriography showing the flow of anomalous origin of the right coronary artery from the main pulmonary artery to the main pulmonary trunk. Collateral artery linked from the left anterior descending artery to the right coronary artery poured into the main pulmonary trunk. White arrow shows collateral artery and the conus branch of the right coronary artery.