Cardiology in the Young

cambridge.org/cty

Characteristics and long-term outcome for congenital left main coronary artery atresia

Etsuko Tsuda[1], Toshikatsu Yagihara[2], Shinya Tsukano[3], Hajime Ichikawa[2] and Soichiro Kitamura[2]

[1]Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Suita, Osaka, Japan; [2]Department of Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Suita, Osaka, Japan and [3]Department of Pediatrics, Niigata City General Hospital, Niigata, Japan

Original Article

Cite this article: Tsuda E, Yagihara T, Tsukano S, Ichikawa H, and Kitamura S (2023) Characteristics and long-term outcome for congenital left main coronary artery atresia. *Cardiology in the Young* **33**: 383–387. doi: 10.1017/S104795112200083X

Received: 4 January 2022
Revised: 21 February 2022
Accepted: 21 February 2022
First published online: 30 March 2022

Keywords:
Left main coronary artery atresia; mitral regurgitation; coronary artery bypass grafting; left main coronary angioplasty; Noonan syndrome

Author for correspondence:
Etsuko Tsuda, Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, 6-1 Kishibe-shinmachi, Suita-shi, Osaka 564-8565, Japan. Tel: +81 6 6170 1070; Fax: +81 6 6170 1782.
E-mail: etsuda@ncvc.go.jp

Abstract

The prevalence of congenital left main coronary artery atresia is very low. We report the characteristics and long-term outcomes of four children with left main coronary artery atresia. Three patients had heart murmurs due to mitral regurgitation at less than 1 year old. Their myocardial ischaemia worsened on exercise with aging. In the fourth patient, hypertrophic cardiomyopathy and Noonan syndrome were suspected at 1 year old. The development of communicating arteries between the conus branch and the left anterior descending artery was detected at 7 years old. The left main coronary artery atresia was confirmed by a selective coronary angiogram at 15 years old. Congenital left main coronary artery atresia could not be diagnosed by two-dimensional echocardiography; however, the left coronary arteries were small. Two patients underwent coronary artery bypass grafting of the left anterior descending artery using the left internal thoracic artery at 3 years and 6 years old, respectively. Two patients had an angioplasty with a cut back at the orifice of the left coronary artery at 2 years old and 17 years old, respectively. Two patients had no cardiac events without medication for more than 30 years after the operation. We must differentiate the diagnosis of left main coronary artery atresia in the small left coronary arteries with mitral regurgitation during the first year. Coronary artery revascularisation and mitral annuloplasty are needed. The long-term outcome of both coronary artery bypass grafting and angioplasty were good. The degree of mitral regurgitation after surgery may affect the prognosis.

The prevalence of congenital left main coronary artery atresia is very low. According to the literature, no more than 60 cases have been reported up until 2017.[1–4] Therefore, it is difficult to diagnose and treat. Further, the reports about the long-term results of more than 10 years are limited.[4] We report the long-term results of four patients whose diagnosis of congenital left main coronary artery atresia was not confirmed at the initial presentation.

Patient 1

The heart murmur in the first patient was detected at 1 year old. He was diagnosed with mitral regurgitation by two-dimensional echocardiography at 3 years old (Table 1). After walking for 20 min at 6 years old, he became pale and had to squat down to rest. ST-T depression in leads V_2–V_4 was detected during the treadmill test. A selective coronary angiogram of the right coronary artery revealed atresia at the orifice of the left coronary artery (Fig 1 left). His height and body weight were 120 cm and 22 kg, respectively. He underwent a coronary artery bypass grafting of the left anterior descending artery using the left internal thoracic artery and mitral annuloplasty. At the age of 11 years old, the left internal thoracic artery graft was patent on the angiograms (Fig 1 middle, right). Myocardial ischaemia was not detected on exercise ^{201}Tl myocardial perfusion imaging. The ST-T depression during the treadmill test improved. On the two-dimensional echocardiogram at 37 years old, his left ventricular end-diastolic dimension and left ventricular ejection fraction were 50 mm and 61%, respectively, and the mitral regurgitation was *trivial*. The diameters of the right coronary artery and left anterior descending artery by two-dimensional echocardiography were 3.3 mm and 2.9 mm, respectively. He has had no cardiac events for 31 years.

Patient 2

The second patient visited our hospital because of mitral regurgitation at 1 year and 6 months old. A heart murmur was detected at 1 year old. Furosemide, spironolactone, and enalapril were given. His left ventricular end-diastolic dimension and left ventricular ejection fraction were 41 mm (117% of normal) and 61%, respectively, and the mitral regurgitation was severe.

Table 1. Characteristics and treatment of congenital left main coronary artery atresia

Patient	1	2	3	4
Age (years)	37	8	37	35
Gender	Male	Male	Female	Male
Co-exist	–	Mental retardation	–	Noonan syndrome, HCM
The initial presentation				
Age	1 year 7 months	3 months	3 months	7 years
Sign	Heart murmur	Heart murmur	Heart murmur	Collateral arteries
Initial diagnosis	MR	MR	MR	
Diagnosis of CLMCAA				
Age	6 years	1 year 6 months	11 months	15 years
Sign and symptom	Squatting after walking	Faintness	Increase of LVDd	Chest pain
Operation	CABG, MAP	CABG, MAP	Angioplasty, MAP	Angioplasty
Age	6 years	3 years	2 years 2 months	17 years
Height and body weight	120 cm, 22 kg	93 cm, 14 kg	84.5 cm, 11 kg	160 cm, 35 kg
Degree of MR after operation	Trivial	Moderate	Slight	Trivial
Medication in the late period	None	ACEI, Diuretics	None	β blocker
		Aspirin		

CLMCAA, congenital left main coronary artery atresia, HCM, hypertrophic cardiomyopathy; MR, mitral regurgitation, CABG, coronary artery bypass grafting, MAP, mitral annuloplasty; LVDd, left ventricular diastolic dimension; ACEI, angiotensin converting enzyme inhibitor.

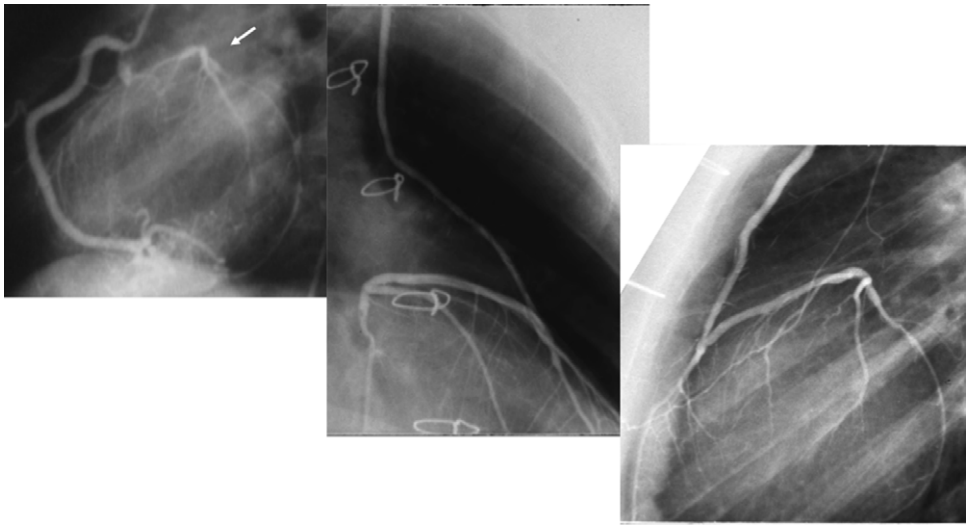

Figure 1. Coronary angiograms before and after the operation (Patient 1) (Left) Right coronary angiogram at the age of 6 years old (Middle and right) Left internal thoracic angiograms at the age of 11 years old.

The diameters of the right coronary artery and left anterior descending artery by 2DE were 1.9 mm and 1.7 mm, respectively (Fig 2 right). When he underwent cardiac catheterisation, he was diagnosed with congenital left main coronary artery atresia (Fig 2 left). A 12-lead electrocardiogram revealed an abnormal Q wave in lead aVL. He had been diagnosed with slight mental retardation. He underwent coronary artery bypass grafting to the left anterior descending artery and mitral annuloplasty at 3 years old in another hospital, because of faintness. His height and body weight at the time of the operation were 94 cm and 14 kg, respectively. The abnormal Q wave in lead aVL disappeared after surgery. The mitral regurgitation remained moderate at 8 years old; however, the left internal thoracic artery graft was patent on the angiograms after the operation.

Patient 3

The third patient visited our hospital at 7 months old, after a heart murmur due to mitral regurgitation was detected at 3 months old. Her left ventricular end-diastolic dimension on a two-dimensional echocardiogram was increased at 11 months old. When she underwent a cardiac catheterisation, severe stenosis of the left main coronary artery was suspected. The atresia at the orifice of the left coronary artery was diagnosed at 2 years old during the operation. The orifice of the left coronary artery existed at just upper 2 mm in the commissure between the left cusp and the non-cusp. Her height and body weight at the operation were 87 cm and 11 kg, respectively. The diameter of the left main trunk was 1.5 mm. A fibrous ridge of the left main trunk was removed. She underwent an angioplasty at

https://doi.org/10.1017/S104795112200083X Published online by Cambridge University Press

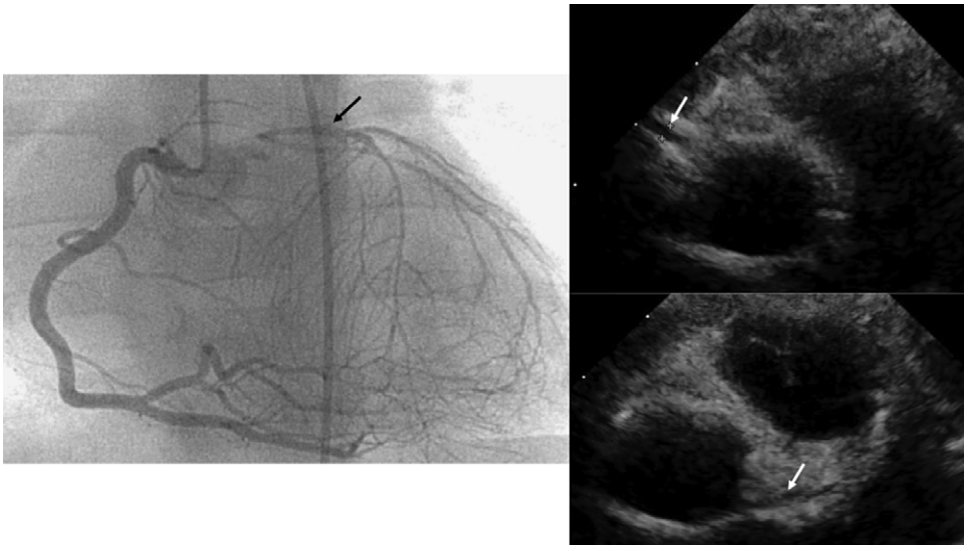

Figure 2. Coronary angiogram and echocardiograms before the operation (Patient 2) (Left) Right coronary angiograms at the age of 1 year and 6 months (Patient 2) (Right upper) Right coronary artery (Right lower) Left coronary artery The diameters of the right coronary artery and the left anterior descending artery were 1.9 mm and 1.7 mm, respectively.

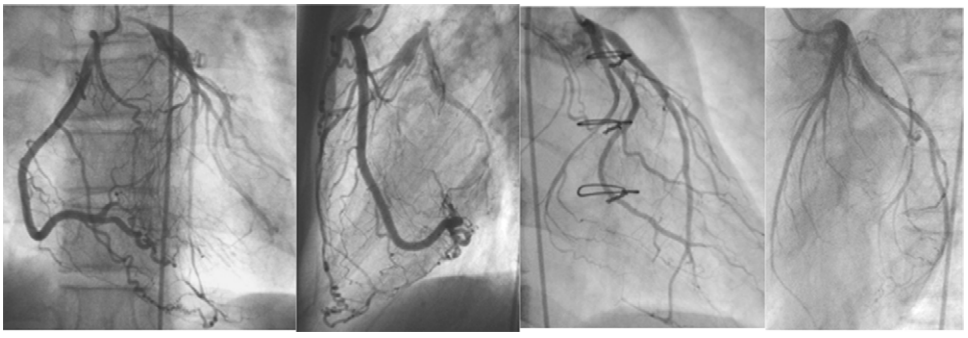

Figure 3. Coronary angiograms before and after the operation (Patient 4) (Left) Right coronary angiograms before the operation (Right) Left coronary angiograms after the operation.

the orifice of the left coronary artery with a cut back and mitral annuloplasty. At the age of 11 years old, the orifice of the left coronary artery was well injected on the coronary angiogram. She delivered two children without any complications in her thirties. In the two-dimensional echocardiography at 37 years old, her left ventricular end-diastolic dimension and left ventricular ejection fraction were 51 mm and 73%, respectively, and the mitral regurgitation was slight. The left ventricular end-diastolic dimension and left atrial dimension were slightly dilated. The diameter of the right coronary artery, left coronary artery, and left anterior descending artery on two-dimensional echocardiography were 3.5 mm, 3.5 mm, and 3.0 mm, respectively.

Patient 4

In the fourth patient, hypertrophic cardiomyopathy and Noonan syndrome were suspected at 1 year old. During the cardiac catheterisation at 7 years old, the communicating arteries between the conus branch and the left anterior descending artery had become well developed at the previous hospital. He sometimes had chest pain during exercise with aging. ST-T depression in leads V_2–V_4 was detected during a treadmill test. He was referred to our hospital. No perfusion defects were detected during the myocardial perfusion imaging with exercise. The diagnosis of left main coronary artery atresia was confirmed in the right coronary angiogram at 15 years old (Fig 3 left). An aneurysm of the left main coronary artery was also detected. The mitral regurgitation detected by two-dimensional echocardiography was trivial. It was also considered that angioplasty of the left coronary artery was possible by CT imaging. He underwent an angioplasty at the orifice of the left coronary artery at 17 years old. The coronary angioplasty was perfomed with a cut back method for membranous atresia. One year after the surgery, collateral arteries disappeared on the coronary angiograms, and the left coronary artery was well injected (Fig 3 right). He had no cardiac events after the surgery up until 35 years old.

Discussion

In patients with mitral regurgitation detected within the first year after birth, mitral regurgitation should be differentiated from secondary mitral regurgitation caused by congenital left main coronary artery atresia.[5–7] Its diagnosis would be difficult, because of its rarity and lack of abnormal findings. The left coronary artery can be detected as normal by two-dimensional echocardiography. The orifice of the left coronary artery seems to be normal at its position on the two-dimensional echocardiogram; however, the left coronary artery is slightly small.[8] The diameter of the right coronary artery is not always dilated on the two-dimensional

https://doi.org/10.1017/S104795112200083X Published online by Cambridge University Press

echocardiogram at the initial presentation. Therefore, it is likely that a diagnosis of atresia of the left coronary artery will be missed. Severe myocardial ischaemia appears depending on an increase in activity with aging. Further examinations should be performed with occurrences such as an increase in the left ventricular end-diastolic dimension with worsening of mitral regurgitation and cardiac events such as chest pain, faintness, and squatting.[1] The stronger the myocardial ischaemia, the earlier the appearance of symptoms. When myocardial ischaemia is speculated in patients with mitral regurgitation, coronary artery abnormalities should be re-evaluated. A coronary angiogram is needed to confirm the left main coronary artery atresia precisely. A selective coronary angiogram can reveal a small left coronary artery when injected retrogradely from the right coronary artery. CT angiography may be convenient for investigating congenital coronary abnormalities.[9,10]

There was a case report about the left main coronary atresia with Noonan syndrome.[11] Further, the coronary aneurysm of the left main coronary artery in the fourth patient may have been related to the characteristics of Noonan syndrome associated with hypertrophic cardiomyopathy.[12] The relationship between coronary abnormalities and Noonan syndrome and its gene abnormalities is very interesting.[11,13]

Our long-term results showed that coronary artery revascularisation of congenital left main coronary artery atresia was useful for both angioplasty and coronary artery bypass grafting.[14] Generally, most of the lumen of the left coronary artery in congenital left main coronary artery atresia reaches just the proximal portion of the left aortic cusp; however, it is often hypoplastic. Therefore, it may be possible to repair it using the cut back method or the patch angioplasty. Further, some methods for a repair of the left main coronary artery and percutaneous transluminal coronary intervention have also been reported.[15,16] To choose the method for the coronary artery revascularisation surgery in each patient, CT imaging to measure the distance between the aorta and the lumen of the left coronary artery may be useful.[5] If an angioplasty of the left main trunk is difficult, coronary artery bypass grafting of the left anterior descending artery is also useful even in children to improve myocardial ischaemia.[5–7,17] Even if there is one graft to the left anterior descending artery without a graft to the left circumflex, their outcome and morbidity could be preserved. The long-term outcome of coronary revascularisation for congenital left main coronary artery atresia was good. However, the degree of mitral regurgitation after surgery may affect the prognosis.

There is a difference in the time of the diagnosis between children and adult patients with congenital left main coronary artery atresia. In most cases of congenital left main coronary artery atresia, the diagnosis is made in childhood. In the child type of congenital left main coronary artery atresia without well-developed collateral arteries, usually the disease is detected by a heart murmur within the first year after birth, which is induced by mitral regurgitation due to myocardial ischaemia. Mitral valve tethering also occurs. The improvement in the degree of mitral regurgitation is also reported with the development of collateral arteries. On the other hand, fatal cardiac events such as an acute myocardial infarction and severe heart failure have been reported in patients with congenital left main coronary artery atresia.[16] An early diagnosis is possible if an appropriate examination is performed with the knowledge of the signs and symptoms of the speculated underlying disease. Further, congenital left main coronary artery atresia in adults is often detected in cases with well-developed collateral arteries from the right coronary artery to the left coronary artery.[9,18,19] Chest pain on effort and arrhythmias occur due to myocardial ischaemia and involvement due to the progression of the disease and atherosclerosis with aging.

Conclusion

In patients with mitral regurgitation within the first year after birth, mitral regurgitation should be differentiated from secondary mitral regurgitation caused by congenital coronary abnormalities. The long-term outcome in the patients with congenital left main coronary artery atresia who underwent either coronary artery bypass grafting or angioplasty was good.

Acknowledgements. None.

Funding. This study received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Conflicts of interest. None.

Ethical approval. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional committee with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

References

1. Musiani A, Cernigliaro C, Maelli D, Gasperis CD. Left main coronary atresia; Literature review and therapeutical considerations. Eur J Cardiothoracic Surg 1997; 11: 505–514.
2. Tanawuttiwat T, O'Nell BP, Schob AH, Alfonso CE. Left main coronary atresia. J Card Surg 2013; 28: 37–46.
3. Koh E, Nakagawa M, Hamada K, et al. Congenital atresia of the left coronary ostium: diagnosis and surgical treatment. Pediatr Cardiol 1989; 10: 159–162.
4. Yajima S, Toda K, Nishi H, et al. Redo coronary bypass grafting for congenital left main coronary atresia: a case report. J Cardio Surg 2017; 12: 26.
5. D'Souza TF, Sameuel BP, Vettukattil JJ, et al. Surgical treatment of neonate with congenital left main coronary atresia. Ann Thorac Surg 2016; 101: 352–325.
6. Ryu SW, Pyo WK, Choi ES, et al. Surgical treatment for left main coronary atresia with significant mitral regurgitation in a 1-year-old child. J Chest Surg. 2021:54: 72–74.
7. Ahmad Z, Haw M, Vettukattil JJ. Congenital mitral regurgitation due to papillary muscle infarction: a case report and approach to evaluation. Eur J Pediatr 2000; 169:621–623.
8. Furuyama H, Kinugawa Y, Nakajima M, et al. Congenital atresia of left main coronary artery followed up for ages as a sequela of Kawasaki disease. Pediatr Int 2010; 52, e131–133.
9. Saedi S, Pouraliakbar HR, Ghaderian H, Saedi T. Congenital atresia of left main coronary artery. Egypt Hear J. 2018; 70: 451–453.
10. Nicol ED, Lyne J, Rubens MB, Padley SG, Ho SY. Left main coronary atresia: a more commonly identified condition after the advent of 64-slice CT coronary angiography? J Nucl Cardiol 2007; 715–718.
11. Ramond F, Duband S, Croisille P, et al. Expanding the cardiac spectrum of Noonan syndrome with RIT1 variant: left main coronary artery atresia causing sudden death. Eur J Med Gene 2017; 60: 299–302.
12. Hakim FA, Gruden JF, Panse PM, Alegria JR. Coronary artery ectasia in an adult Noonan syndrome detected coronary CT angiography. Heart Lung Circ 2013; 22: 1051–1053.
13. Ly R, Soulat G, Iserin L, Ladouceur M. Coronary artery disease in adults with Noonan syndrome: case series and literature review. Arc Cardivasc Dis 2021; 114: 598–605.

https://doi.org/10.1017/S104795112200083X Published online by Cambridge University Press

14. Kitamura S. Pediatric coronary artery bypass surgery for congenital heart disease. Ann Thorac Surg 2015; 106: 1570–1577.
15. Sunagawa M, Shimabukuro T, Kado H, Ushonohama H, Ohta T. Congenital atresia of the left main coronary artery: successful surgical treatment (myocardial revascularization and mitral valve repair). Eur J Pediatr 2005; 164: 461–462.
16. Yin T, Zhong X, Tao L, Hu T. Revascularization by percutaneous coronary intervention in a case of left main coronary artery atresia. Cath Cardiovasc Inter 2021:98: 1343–1348.
17. Gebauer R, Cerny S, Vojtovic P, Tax P. Congenital atresia of the left coronary artery–myocardial revascularization in two children. Interactive Cardiovasc Thoracic Surg 2008; 7: 1174–1175.
18. Sohn SY, Jang GY, Choi BM. Congenital atresia of the left main coronary artery in an infant. J Zhejiang Univ Sci B. 2010; 11: 539–541.
19. Kinoshita H, Murata S, Kazuno K, Sato H. A rare case of mitral regurgitation with congenital left main coronary artery atresia. Jpn J Cardiovasc Surg 2018; 47: 18–21.

https://doi.org/10.1017/S104795112200083X Published online by Cambridge University Press