CASE REPORT

Congenitally corrected transposition of the great arteries in an 83-year-old asymptomatic patient: description and literature review

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SUMMARY

We describe the case of an 83-year-old asymptomatic man followed in our centre. Transoesophageal echocardiography disclosed congenitally corrected transposition of great arteries (CCTGA) with no associated anomalies and only mild aortic regurgitation. Cardiac MR confirmed the diagnosis and revealed preserved systemic ventricle systolic function with a normal perfusional pathway. This report is a demonstration that CCTGA without associated anomalies can reach older life in an asymptomatic condition. This is the oldest asymptomatic living patient with CCTGA ever described.

BACKGROUND

Congenitally corrected transposition of great arteries (CCTGA) is a rare congenital heart disease with a poor prognosis.^{1–3} The physiological and anatomic characteristics of an aged asymptomatic patient is described.

CASE PRESENTATION

An 83-year-old asymptomatic man was referred to our centre for cardiology consultation after clinical examination revealed a 3/6 holosystolic and 2/6 decrescendo diastolic murmur at the left sternal border. A chest radiograph showed a normal cardiac index and an egg-shaped ventricle; the liver shadow and stomach bubble were located normally.

The ECG showed first-degree atrioventricular block (AVB), left axis deviation. Q waves were present in lead V1 and absent in leads V5, V6. The inferior leads showed abnormal Q inferior waves that mimicked inferior myocardial infarction and T waves were negative in DI, aVL (figure 1).

Owing to a poor acoustic window we performed transoesophageal echocardiography that showed the aorta arising from the morphological right ventricle (RV), which was identified by the three leaflet tricuspid valve that inserted more apically than the mitral valve. The pulmonary trunk, identified by its bifurcation, arose from the morphological left ventricle (LV). The systemic RV was dilated, hypertrophic, and the systolic RV function was normal (ejection fraction 55%). There was mild atrioventricular (tricuspid) valve regurgitation. The pulmonary ventricle (LV) showed normal function and there was a mild atrioventricular (mitral) valve regurgitation. There was a mild aortic regurgitation but no pulmonary valve stenosis. We found a patent foramen ovalis with left to right shunt.

Cardiovascular MR (CMR) confirmed the diagnosis, assessing more precisely RV systolic function that was substantially preserved (ejection fraction 56%; figures 2 and 3).

Contrast-enhanced CMR with myocardial late gadolinium enhancement study did not show any myocardial area of delayed hyperenhancement suggestive of structural damage (fibrosis).

Figure 1 ECG showing the common atrioventricular conduction defect and abnormal Q waves that mimic inferior myocardial infarction.

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Figure 2 Axial spin-echo MRI demonstrating a left-anterior placed aorta, with left coronary artery visible and a right-posterior placed pulmonary artery.

Figure 3 Gradient-echo MRI in the four chambers view illustrates the moderator band and the heavy trabeculations and the more apically septal attachment of the tricuspid valve relative to the mitral valve at the level of the left ventricle (LV; morphological right ventricle (RV)).

At follow-up, the patient experienced persistent atrial fibrillation, well prevented with amiodaron and warfarin. A few years after the diagnosis the mild aortic regurgitation became moderate, but the patient did not experience any episode of heart failure, and had normal functioning of the systemic RV.

DISCUSSION

The CCTGA is a rare congenital heart disease often diagnosed in adult age.³ This finding can be explained by the fact that echocardiography was not available in the era involved and patients with CCTGA without other anomalies did not require medical attention in childhood.

We reported the oldest asymptomatic living man having this rare disease, with normal systolic systemic RV function and no significant valvular regurgitation (table 1).

Only nine living patients with congenitally corrected transposition of the great vessels older than 70 years have been reported in the literature. Our patient's survival is the longest, without history of cardiac failure symptoms, ever described. Table 1 shows that almost all patients have no advanced AVB. In fact, one of the most common causes of heart failure is the bradycardia requiring pacemaker implant, which is so frequent in this disease. The mechanism proposed is the excessive length of the abnormally located atrioventricular bundle and its chronic stress during closure of the atrioventricular right (mitral) valve causing fibrosis and damage. As to tricuspid regurgitation, it is not clear if this condition causes RV dysfunction or if ventricular failure determines the valve incompetence. Each scenario can probably play various roles in different patients. Hornung *et al*¹¹ show another possible cause of systemic RV failure: the inadequate coronary arterial flow is not sufficient to satisfy hypertrophied myocardium oxygen request, developing myocardial structural damage (fibrosis) despite patent coronary arteries and subsequent chronic irreversible right ventricular dysfunction.¹¹ In our patient, myocardial late gadolinium enhancement CMR revealed a normal pathway; this can explain the normal life of the patient. Another hypothesis is RV remodelling with unfavourable shape to sustain systemic pressures lifelong, which becomes larger and longer due to the effect of Laplace law until the irreversible dilation causes heart failure.

In conclusion, the prognosis of CCTGA without associated cardiac anomalies depends on the appearance of anatomical tricuspid regurgitation and subsequent development of systemic (morphological right) ventricular dysfunction and advanced heart block.

CMR clearly depicted all morphological findings of CCTGA and assessed, precisely, right ventricular function, confirming

Table 1 Clinical variables in reports of patients older than 70 years with CCTGA

	Sloth <i>et al</i> ⁴	Kowalik <i>et al</i> ⁵	Roffi <i>et al</i> ⁶	Pézarid <i>et al</i> ⁷	Pézarid <i>et al</i> ⁷	Misumi <i>et al</i> ⁸	Crepaz <i>et al</i> ⁹	Orchard <i>et al</i> ¹⁰
Age	76	72	80	72	80	81	75	82
Gender	F	M	F	M	F	M	M	F
CHF	N	Y	Y	Y	Y	Y	N	N
RV dysfunction	N	Y	Y	Y	Y	Y	N	N
Significant TR	N	Y	Y	Y	Y	Y	N	N
Pacemaker	Y	N	N	N	N	N	N	N
Arrhythmia	SR	Y	SR	AF	SR	SR	VE	SR
Significant aortic R	N	N	Y	Y	N	N	N	N

AF, atrial fibrillation; CCTGA, congenitally corrected transposition of great arteries; CHF, congestive heart failure; RV, right ventricle; SR, sinus rhythm; TR, tricuspid regurgitation; VE, ventricular ectopy.

itself as a very important non-invasive imaging technique for the morphological and functional evaluation of complex congenital cardiovascular diseases especially in adults.^{12 13} CMR may be optimal as a confirmation study also because it overcomes some echocardiographic limitations (insufficient acoustic window, difficult visualisation of right ventricular chamber), and especially because it allows the most accurate measurement of systemic ventricular function parameters,¹⁴ providing additional information to correctly define prognosis (late enhancement study).¹⁵

We believe this to be the oldest asymptomatic patient with corrected transposition of the great arteries without associated anomalies whose case was reported in the literature and whose diagnosis was established *in vivo*.

Learning points

- Congenitally corrected transposition of great arteries is a rare congenital heart disease normally with a poor prognosis because of several associated anomalies.
- The prognosis in patients without anomalies is varied and can be acquired atrioventricular block, atrioventricular tricuspidal (systemic) valve regurgitation or right ventricle dysfunction.
- Cardiovascular MR is the gold standard for anatomical and functional diagnosis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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