A case report of paroxysmal complete atrioventricular block in a patient with dextrocardia and repaired tetralogy of Fallot

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Background
Some adults suffer sudden cardiac death after previous surgical repair of tetralogy of Fallot (TOF), and in such cases, ventricular tachycardia is believed to be the most frequent cause of death. However, we report a case of cardiac arrest due to paroxysmal complete atrioventricular block in an adult with dextrocardia and repaired TOF.

Case summary
A 49-year-old woman with dextrocardia and a history of surgical treatment for TOF lost consciousness three times. A previously implanted loop recorder showed a 60-second cardiac arrest, and complete atrioventricular block was diagnosed. An electrophysiological study showed prolongation of the His-ventricular interval but no ventricular tachycardia. A dual chamber pacemaker was implanted, and there has been no recurrence of syncope in the 23 months since implantation.

Discussion
There is little evidence for paroxysmal complete atrioventricular block in patients with repaired TOF. This case suggests that paroxysmal complete atrioventricular block can occur late after surgical repair of TOF, and research needs to elucidate whether it is the cause of sudden cardiac death in some patients with TOF.

Keywords
Case report • tetralogy of Fallot • sudden cardiac death • paroxysmal complete atrioventricular block • dextrocardia

ESC Curriculum
5.7 Bradycardia • 5.9 Pacemakers • 9.7 Adult congenital heart disease

Learning points
- Sudden death is one of the most critical late postoperative complications in patients with tetralogy of Fallot (TOF).
- Ventricular tachycardia is considered to be the main cause of sudden death in these patients.
- This case report suggests that paroxysmal complete atrioventricular block may be the cause of sudden death in some patients with repaired TOF.

Introduction
The long-term outcome of patients with repaired tetralogy of Fallot (TOF) has improved over time. However, sudden cardiac death remains the most critical complication late after surgical repair of TOF, and ventricular tachycardia is well known as the main cause. On the other hand, little information is available about atrioventricular block (AVB) late after repair of TOF. Here, we report a case of cardiac arrest due to paroxysmal complete AVB in a patient after previous repair of TOF and suggest that complete AVB may be the cause of sudden cardiac death in some cases.
Timeline

| Event                                      | Date                        |
|--------------------------------------------|-----------------------------|
| Tetralogy of Fallot repaired by the        | At age 7 (in 1978):         |
| transannular patch approach                 |                             |
| First syncope in the kitchen                | 11 months before current    |
| admission:                                  | admission:                  |
| Second syncope in the bathroom. Implantable|                             |
| loop recorder (ILR) was implanted.          | 9 months before current     |
| admission:                                  | admission:                  |
| Third syncope during dinner                 |                             |
| ILR showed 60-second cardiac arrest         | Day of admission:           |
| with regular P waves.                       |                             |
| Dual chamber pacemaker was implanted.       | Day 9 of admission:         |
| Patient was discharged.                     | Day 22 of admission:        |

Case presentation

The patient was a 49-year-old woman with a history of dextrocardia, situs inversus, and surgical repair of TOF. At the age of 2, she underwent palliative shunt surgery for TOF. At the age of 7, TOF was repaired by the transannular patch approach. It was presumed that ventricular septal defect (VSD) had been repaired by Teflon felt patch. When she was in her twenties, she often felt palpitations and dyspnoea and was diagnosed with panic disorder. She had chronic hepatitis C and achieved sustained virological response with glecaprevir and pibrentasvir treatment at the age of 48 (1 month after the first admission). She was prescribed oestradiol, dydrogesterone for menopausal disorders, and escitalopram, alprazolam for panic disorder. At the age of 48, she fainted for the first time in the kitchen. When she recovered consciousness, she was lying on the floor and realized that she had suffered a head injury, but she did not go to hospital. Two months later, she presented to the emergency department of our hospital because of a recurrence of syncope in the bathroom. Before the syncope, she had experienced palpitation and dyspnoea. At the time of examination, she was conscious and had no complaints. A 12-lead electrocardiogram (ECG) showed no significant changes; results of other tests were normal, including electrolytes and cardiac enzymes and findings of a two-dimensional echocardiogram. An implantable loop recorder (ILR) was implanted to evaluate the cause of syncope. Nine months later, she briefly became unconscious while having dinner with friends but then regained consciousness. Three days later, she visited our hospital. The ILR showed that the syncope had been caused by a 60-second complete atrioventricular block (Figure 1). The patient’s blood pressure was 113/75 mm Hg, her heart rate was 84 bpm, and her heart rhythm was regular. She had no signs of heart failure, such as congested neck veins, pulmonary adventitious sound, heart murmur, or leg oedema. A 12-lead ECG showed prolonged (PR) the time from the onset of the P wave to the start of the QRS complex and right bundle branch block (Figure 2A). Chest X-ray confirmed dextrocardia but showed no cardiac enlargement (Figure 2B). Laboratory tests found no abnormalities of electrolytes, thyroid hormones, or brain natriuretic peptide levels. The echocardiography showed left ventricular ejection fraction (LVEF) was 64%, no asynergy motion and no enlargement of left ventricle (LV). And it also found normal size and motion of right ventricle (RV) had moderate pulmonary regurgitation. The detailed ILR data showed regular P waves during a significant cardiac arrest (Figure 3). On the basis of the findings, we diagnosed paroxysmal AVB. Cardiac computed tomographic (CT) angiography revealed no stenosis in the coronary artery. In the head-up tilt test, the vasovagal reflex was induced without bradycardia (blood pressure, 67/33 mm Hg; heart rate, 104 bpm).

An electrophysiological study found that the His-ventricular interval was prolonged (66 milliseconds). Ventricular tachycardia was not induced by programmed ventricular stimulation (burst pacing and up to three extra stimuli) at two right ventricular sites (apex and outflow; Figure 4). A dual chamber pacemaker was implanted on the 9th day of admission (Figure 5). The patient was discharged on the 22nd day of admission and has not experienced any symptoms, including syncope, in the 23 months since then.

Discussion

TOF is one of the most common congenital heart diseases (CHD) and accounts for 3.5–10% of cases. However, TOF with dextrocardia is rare and accounts for 0.8–1.4% of cases of CHD. To our knowledge,
Figure 2. (A) Twelve-lead electrocardiogram showing sinus rhythm and three bundle blocks (first-degree atrioventricular block, complete right bundle branch block, and left anterior hemiblock) (B) chest X-ray showing dextrocardia.
no report has described complete AVB in a patient with repaired TOF and dextrocardia.

The long-term outcome after surgical repair of TOF has improved over time, and the 30-year survival rate had reached almost 90% by the late 1990s. However, sudden cardiac death is still recognized as a late complication. Ventricular tachycardia is the main cause of sudden death in patients with TOF; one study found that approximately 12% of patients experienced ventricular tachycardia late after TOF surgery and 8% of them died suddenly more than 35 years after the procedure.

The prevalence of paroxysmal AVB in patients with TOF is unknown. Hokanson et al. showed that transient complete AVB after the third operative day was a significant risk factor for sudden death, probably because of recurrent AVB. Some reports also found that transient complete AVB is associated with late complete AVB and that sudden death can occur in the presence of right bundle branch block and left anterior hemiblock in patients after surgical repair of TOF.

Paroxysmal AVB appears to be caused by a distal conduction disorder, such as bundle branch block. Vagal effects may trigger cardiac arrest. In general, the impaired His-Purkinje system cannot spontaneously depolarize and becomes less responsive to subsequent impulses because of sodium channel inactivation. In the present case, the vasovagal reflex was induced by the head-up tilt test. Therefore, the vagal reflex may have triggered the onset of AVB and asystole, and the impaired His-Purkinje system may have prolonged the duration of the block.

Nakazawa et al. reported that postoperative complete AVB was seen in patients with a perimembranous VSD. In our patient, we assume that a distal conduction disorder occurred because of the earlier surgical procedures (especially with VSD) and age-related changes in myocardial tissues and that this disorder led to the paroxysmal complete AVB late after surgical repair of TOF. Delarue et al. showed the case report of high degree atrioventricular block (right bundle branch block and left anterior hemiblock) and recurrent syncope in the patient with repaired TOF. They also implanted a pacemaker in the patient and improved the symptom. More cases of patients late after surgical repair of TOF seem to experience paroxysmal AVB due to the additional age-related changes in the conduction system on top of congenital (including surgical) predisposition to conduction abnormalities. To investigate the prevalence of paroxysmal AVB in patients with TOF would be warranted.

To the best of our knowledge, the current report is the first to describe cardiac arrest due to paroxysmal complete AVB late after repair of TOF in a patient with dextrocardia. Further research is needed to determine whether paroxysmal complete AVB is responsible for sudden cardiac death in patients with repaired TOF.

Figure 3 Record from the implantable loop recorder showing the complete atrioventricular block, which is a 5.5 times magnification of Figure 1. ▼: onset of the pause with third-degree atrioventricular block. ▲: regular P waves during the pause.
A case report of paroxysmal complete atrioventricular block

Lead author biography

Takeshi Fujita graduated from Osaka University and received the MD degree in 2017. He was working at Department of Cardiology in Higashi-Osaka Medical Center from 2020 to 2021.

Supplementary material

Supplementary material is available at European Heart Journal – Case Reports.

Slide sets: A fully edited slide set detailing these cases 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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