Infective endocarditis in an adult with unrepaired corrected transposition

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Abstract
We report a case of a 28-year-old man with unrepaired congenitally corrected transposition of the great arteries, ventricular septal defect, and pulmonary stenosis who presented with septic shock due to infective endocarditis by Abiotrophia defectiva. The cardiac catheterization had the risk of vegetation scattering. Without invasive hemodynamic assessment, the degree of pulmonary stenosis and left ventricle preparation as a systemic ventricle could not be accurately determined, making surgical planning difficult. We chose a staged approach with pulmonary valve replacement first for source control, followed by a more definitive operation following recovery from endocarditis.

Keywords
Adult congenital heart disease, endocarditis, great vessels anomaly, infection

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Introduction
Treatment of infective endocarditis (IE) in septic shock is associated with a high in-hospital mortality rate (approximately 80%). Furthermore, there was no guideline determining the appropriate treatment strategy in adult patients with unrepaired congenital heart disease without evaluating preoperative hemodynamics. Surgical treatment of IE in patients with unrepaired congenitally corrected transposition of the great arteries (ccTGA), ventricular septal defect (VSD), and pulmonary stenosis (PS) requires considering specific parameters, including pulmonary hypertension, decreased left-sided morphological right ventricular (mRV) function, and left-sided morphological tricuspid regurgitation (mTR) degree. We report an adult with unrepaired ccTGA in septic shock due to IE who was successfully treated using two-stage repair.

Case report
Written informed consent for the use of patient records and publication of the article was obtained from the patient. A 28-year-old man was referred from a local hospital following septic shock due to IE. Although he was diagnosed with ccTGA (L-TGA), VSD, and valvular and sub-valvular PS after birth, he was lost to follow-up after 12 years of age before intracardiac repair.

One week before admission, he developed fever of more than 38 degrees, palpitations, and dyspnea on exertion. On arrival, his sequential organ failure assessment score was nine with epinephrine (0.05 µg/kg/min) and norepinephrine (0.15 µg/kg/min) administration, respiratory distress (the ratio of partial pressure arterial oxygen and fraction of inspired oxygen < 200 with mechanical ventilation), and low platelet count (63,000/mm²). Echocardiography revealed a mobile vegetation of 26 mm in diameter at the pulmonary valve (Figure 1) with 4.4 m/s peak velocity, severely reduced biventricular function with 30% mRV ejection fraction, and moderate mTR. Computed tomography showed multiple septic pulmonary emboli (Figure 1) and no cerebrovascular accident. IE did not improve despite broad antimicrobial therapy using vancomycin and gentamycin; total leucocyte count (13,000–15,000/mm³) and C-reactive protein (CRP) (12–16 mg/dL) remained high for 3 days. Follow-up echocardiography showed disappearance of a small friable portion of the vegetation suggesting recurrence of pulmonary septic...
embolism. Cardiac catheterization for pulmonary hypertension and Qp/Qs evaluation was precluded, owing to the risk of vegetation scattering.

Three days post-admission, we performed palliative surgery. Median sternotomy and aortic and bicaival venous cannulation were performed with antegrade cardioplegia delivery. The pulmonary artery (PA) trunk was opened, revealing a vegetation in the pulmonary outflow (Figure 2). We resected the pulmonary valve and vegetation en bloc and debrided the vegetation attached to the sub-pulmonary conus and upper margin of the VSD. The diameter of pulmonary was 20 mm. Pulmonary valve replacement (PVR) was performed using a 19 mm Epic valve (Abbott, California, USA) sutured to the native PA trunk 1 cm distal from the pulmonary valve annulus. After cardiopulmonary bypass was weaned off, Qp/Qs was 1.6 and mean PA pressure was 32 mmHg; hence, PA banding with 7 mm diameter was added. The mean PA pressure decreased to 25 mmHg. Postoperative electrocardiogram showed a regular sinus rhythm. He was extubated on postoperative day (POD) 3.

*Abiotrophia defectiva* was identified by the vegetation culture and 16S rRNA analysis. We performed tooth extraction and antibiotic therapy (12 g/day ampicillin for 6 weeks

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**Figure 1.** Preoperative findings. (a and b) Transesophageal echocardiography and (c) enhanced cardiac computed tomography reveal pulmonary valve vegetation. Chest CT shows multiple septic emboli (d). Dotted area indicates vegetation. mLV: right-sided morphological left ventricle; mRV: left-sided morphological right ventricle; Ao: aorta; PA: pulmonary artery.

**Figure 2.** Surgical view of the pulmonary valve through pulmonary arteriotomy. A large vegetation is attached to the leaflet. Asc.Ao: ascending aorta; PA: pulmonary arteriotomy.
and 3 mg/kg/day gentamicin for 2 weeks). After initiating heart failure medications, he was discharged home at POD 28. Ten months later, echocardiography showed improved mRV ejection fraction of 54%, moderate-to-severe mTR, and 3.7 m/s peak velocity at PA banding, suggesting low PA pressure. However, cardiac magnetic resonance imaging showed 40% right-sided morphological left ventricular (mLV) ejection fraction, and cardiac catheterization revealed 19 mmHg mLV end-diastolic pressure.

One year postoperatively, he underwent physiological repair, including VSD closure, tricuspid valve replacement with a 27 mm ATS standard mitral valve (Medtronic, Minneapolis, USA), and PA de-banding. He was extubated on POD 2 and discharged home without any complications on POD 11. Two years postoperatively, his condition was satisfactory without recurrence of endocarditis and heart failure.

Discussion
The best practices for IE management in unrepaired ccTGA remain unknown, and the data are controversial due to the rarity of the cases. We presented an adult case of septic shock owing to IE with unrepaired ccTGA, VSD, and PS. It was unclear whether physiological or anatomical repair was better for this patient because of unknown pulmonary pressure and reduced biventricular function.

Although the patient required urgent surgery, we could not perform cardiac catheterization for hemodynamic evaluation owing to the risk of vegetation scattering. PS possibly allowed prophylactic PA banding that prepared the mLV to the systemic ventricle. There were four treatment choices: (1) physiological repair or (2) anatomical repair by (a) staged repair or (b) one-stage repair. Anatomical repair has better long-term prognosis than physiological repair. Several studies reported young adult cases with ccTGA, VSD, PS, and reduced mRV function undergoing anatomical repair.

Then, we performed the palliative procedures: vegetectomy, PVR, and PA banding. The indication and degree of PA banding should be carefully determined, as it may lead to postoperative biventricular failure. Despite receiving heart failure medications, impaired mLV function and high mLV end-diastolic pressure remained. Anatomical repair was not suitable with reference to the criteria by Ibrahimiye et al. Subsequently, the patient underwent physiological repair.

Abiotrophia defectiva is rare as a causative bacteria of IE and an aggressive organism that is difficult to treat in IE, with a treatment failure rate of up to 40%. Embolization occurs in up to one-third of patients with A. defectiva, and most deaths occur due to refractory heart failure or major systemic emboli. We chose a long-time interval between the first palliation and the intracardiac repair not only for heart failure treatment but also for observation for recurrence of infection.

Conclusion
We reported a rare case of unrepaired ccTGA with septic shock due to IE, in which preoperative hemodynamics could not be evaluated. We performed a staged repair and obtained a satisfactory outcome. Close follow-up is recommended in such cases after a physiological repair and the infection due to A. defectiva.

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Ethical approval
Ethical approval to report this case was obtained from our institutional review board (approval number was 16105 approved on 2 November 2016).

Informed consent
Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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