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Total cavopulmonary connection for right ventricular endomyocardial fibrosis

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Abstract. Endomyocardial fibrosis is an endemic problem in tropical countries and is characterised by ventricular cavity obliteration, decreased ventricular compliance, and atrioventricular valve regurgitation. We report on a patient with right ventricular endomyocardial fibrosis resulting in obliteration of the cavity and tricuspid regurgitation treated successfully by total cavopulmonary connection and exclusion of the right ventricle. [Eur J Cardio-thorac Surg (1992) 6:391–392]

Key words: Endomyocardial fibrosis – Total cavopulmonary connection

Total cavopulmonary connection (TCPC) has been utilised in many severe congenital malformations where the right side of the heart has to be bypassed [2, 9, 11]. We report a case of severe right ventricular obliteration by endomyocardial fibrosis (EMF) with severe tricuspid valve regurgitation treated by total cavopulmonary connection and exclusion of the right ventricle (RV).

Case report

A 25 year old female patient was evaluated for severe tricuspid regurgitation with engorged neck veins, ascites, pitting edema, and NYHA Class III symptoms. The symptoms started 3 years prior to the present admission and had been progressive. She underwent cardiac catheterisation and right ventricular muscle biopsy. Angiography revealed obliteration of the right ventricular cavity with grade IV tricuspid regurgitation and a dilated right atrium. The right atrial pressure was a mean of 18 mmHg, the RV was 25/15 mmHg (mean 16 mmHg). Pulmonary artery pressure was 21/10 mmHg (mean 19). The pulmonary arteriolar resistance was computed to be 472 dynes/cm². The pulmonary capillary wedge pressure was 9 mmHg (mean). Muscle biopsy showed normal muscle. A MRI study was inconclusive. The RV obliteration was angiographically highly suggestive but not typical of EMF (Fig. 1).

At operation, she was found to have a grossly enlarged right atrium, a small RV, dimpled across the outflow tract and severe tricuspid valve regurgitation. High superior vena cava cannulation was performed and on bypass, right atriotomy revealed the tricuspid valve apparatus and cusps to be involved in a fibrotic process which obliterated most of the cavity of the right ventricle extending from the inflow to the pulmonary annulus. A frozen section of a representative area showed histology compatible with EMF, showing fibrosis infiltrating muscle layers. It was elected to exclude the RV by closing the tricuspid valve with a pericardial patch and detaching the main pulmonary artery and closing the RV outflow tract. The superior vena cava was transected and the proximal part was anastomosed to the right branch of the pulmonary artery and the cavo atrial junction was closed. The main pulmonary artery was anastomosed to the right atrial appendage after tailoring it down to a near tubular conduit, draining the inferior vena cava through the body of the right atrium. She was weaned off bypass in sinus rhythm. The patient was extubated in the immediate postoperative period and she maintained good cardiac output and gas exchange.

She was transferred from the ICU on the 1st postoperative day. Chest tubes were selectively left until until the 7th postoperative day and a vigorous diuresis was maintained. Ascites improved and transesophageal echocardiography on the 8th postoperative day showed unrestricted cavopulmonary flow and diminished right ventricular size with no flow. Left ventricular function was good. The patient was discharged home 3 weeks after surgery.

Discussion

EMF, first described by Davies in Uganda (1948), is an endemic problem in tropical countries and rarely seen in a Caucasian population. This disorder is characterised by fibrotic thickening of the endocardium infiltrating the myocardium associated with partial obliteration of one
or both ventricular cavities resulting in decreased ventricular distensibility and impairment of filling [8].

The basic fibrotic process may involve either ventricle individually or both ventricles together. The isolated right ventricular form of the disease involves characteristically only the inflow portion and papillary muscles. This is a pattern that is more commonly observed in India [10]. In Uganda, biventricular or left ventricle involvement is more common. The disease can sometimes start in one ventricle and later involve the other [7]. The fibrotic process usually starts in the apical region and extends to the inflow region. Involvement of the valve apparatus causing regurgitation of either atrioventricular valve is also a common feature. The diastolic restriction associated with severe atrioventricular regurgitation leads to circulatory congestion and forms the main indication for surgery [7, 8]. Patients are considered to be in a burnt-out phase (chronic), when their history of symptoms is greater than 2 years.

Echocardiography nowadays is the mainstay in diagnosis [6]. Right ventricular angiograms in fully established EMF show obliteration of the RV apex, dilatation of RV outflow tract, tricuspid regurgitation and an enlarged right atrium. However, atypical patterns of early EMF with only trabeculation abnormalities on angiograms have been proven by histology [13]. In the present case even though the tricuspid regurgitation and obliteration of the RV cavity in the inflow region is clearly seen, the characteristic apical obliteration is replaced by trabecular abnormalities. However, in view of the histology, the diagnosis of EMF is acceptable.

The first surgical attempt at decorticating the EMF was carried out by Dubost in 1971 and since then, many series of surgical corrections have been published [4, 5, 14]. The rationale of the surgical approach is centered around decorticating the ventricular endocardial fibrous tissue to improve ventricular compliance and atrioventricular valve replacement for correction of regurgitation. Valve replacement in this condition has its unique problems, sometime tissue valves cannot be used because of space limitation within the obliterated cavities. Mechanical valves in the tricuspid position are extremely difficult to manage and are liable to thrombosis despite adequate anticoagulation. There has been one report of cavopulmonary shunt (Glenn’s operation) being done in young patients for palliation [15].

In the present case, we found the RV cavity of inadequate size and function to be kept in the circulation. It was also felt that following decorticating, function may deteriorate further and the involvement of the tricuspid valve was extensive enough to warrant replacement. Therefore it seemed logical to exclude the right ventricle [1]. The lack of pulmonary vascular disease, as evidenced by the low mean pulmonary arterial pressure (19 mmHg) and a significantly low pulmonary arterial resistance (472 dynes/cm²), coupled with a right atrial pressure of 18 mmHg, was in accordance with the indication for a Fontan-type correction. We were also encouraged by a report of RV exclusion in a patient with cardiac tumour corrected by the Fontan procedure [3]. We carried out TCPC because of its documented hemodynamic advantage [12].

We report this case as there have been no previous reports of this procedure in the treatment of EMF. We propose that TCPC is a viable option in certain cases of right ventricular EMF where either residual cavity size or function would invalidate standard forms of surgical therapy particularly when the tricuspid valve is extensively involved and needs replacement.

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References


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