Peer reviewed article

Ebstein's anomaly: one and a half ventricular repair

Antonio F. Corno, Pierre-Guy Chassot, Maurice Payot, Nicole Sekarski, Piergiorgio Tozzi, Ludwig K. von Segesser

Summary

Patients with Ebstein's anomaly can present after childhood or adolescence with cyanosis, arrhythmias, severe right ventricular dysfunction and frequently with left ventricular dysfunction secondary to the prolonged cyanosis and to the right ventricular interference. At this point conventional repair is accompanied by elevated mortality and morbidity and poor functional results. We report our experience with three patients (8, 16 and 35 years of age) with Ebstein's anomaly, very dilated right atrium, severe tricuspid valve regurgitation (4/4), bi-directional shunt through an atrial septal defect and reduced left ventricular function (mean ejection fraction = 58%, mean shortening fraction = 25%). All underwent one and a half ventricular repair consisting of closure of the atrial septal defect, tricuspid repair with reduction of the atrialised portion of the right ventricle and end-to-side anastomosis of the superior vena cava to the right pulmonary artery. All patients survived, with a mean follow-up of 33 months. In all there was complete regression of the cyanosis and of the signs of heart failure. Postoperative echocardiography showed reduced degree of tricuspid regurgitation (2/4) and improvement of the left ventricular function (mean ejection fraction = 77%, mean shortening fraction = 40%). In patients with Ebstein's anomaly referred late for surgery with severely compromised right ventricular function or even with reduced biventricular function, the presence of a relatively hypoplastic and/or malfunctioning right ventricular chamber inadequate to sustain the entire systemic venous return but capable of managing part of the systemic venous return, permits a one and a half ventricular repair with good functional results.

Key words: congenital heart defects; congenital heart surgery; cyanosis; Ebstein's anomaly; one and a half ventricular repair; tricuspid valve

Introduction

Ebstein's anomaly of the tricuspid valve is a congenital heart defect consisting of downward displacement of the septal and posterior leaflets into the right ventricle, resulting in the formation of an atrialised portion of the right ventricle [1]. This congenital heart defect is frequently associated with the presence of an atrial septal defect and with tricuspid valve regurgitation and, more rarely, stenosis.

The age of presentation reflects the degree of tricuspid and right ventricular abnormality. Ebstein's anomaly can present with signs of severe cyanosis and right heart failure from early infancy, perhaps requiring very early surgical treatment [2].

In most cases, patients with Ebstein's anomaly present after infancy with a combination of moderate cyanosis and right heart failure and at this point conventional surgery is considered [3].

Late referral, after childhood or adolescence is a more rare occurrence. At this age patients often present with cyanosis, arrhythmias, severe right ventricular dysfunction, frequently accompanied by left ventricular dysfunction secondary to the long standing cyanosis and to the interference in right ventricular dysfunction. At this point the isolated conventional repair, suggested by Carpentier based upon the type of anatomic anomaly [1] is either impossible or accompanied by heightened mortality and morbidity and poor functional results.

We report our experience with three patients with Ebstein's anomaly surgically treated with one and a half ventricular type of repair.

No financial support to declare.

Patients and methods

Three consecutive patients, respectively 8, 16 and 35 years of age, presented with cyanosis and heart failure. In all of them echocardiography confirmed the presence of Ebstein's anomaly (figure 1) with a very dilated right atrium (figure 2), severe tricuspid valve regurgitation (figure 3), bi-directional shunt through an atrial septal defect (figure 4) and left ventricular function with a mean ejection fraction = 58% and mean shortening fraction = 25%.

All of them underwent one and a half ventricular type of surgical repair consisting of patch closure of the atrial septal defect, repair of the tricuspid valve with reduction of the atrialised portion of the right ventricle and end-to-side anastomosis of the superior vena cava to the right pulmonary artery (= bi-directional Glenn).

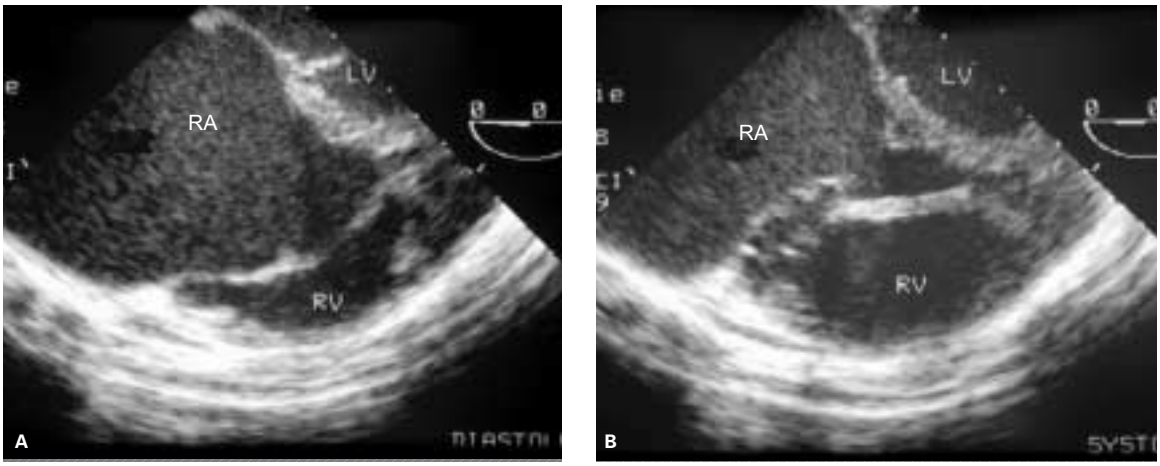

Figure 1

Pre-operative transoesophageal echocardiography, showing the typical feature of Ebstein's anomaly in diastole (A) and systole (B) RA = right atrium, RV = right ventricle.

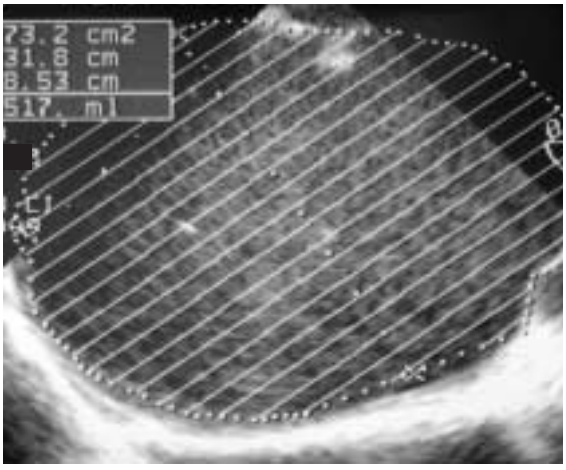

Figure 2

Pre-operative transoesophageal echocardiography, showing a very dilated right atrium.

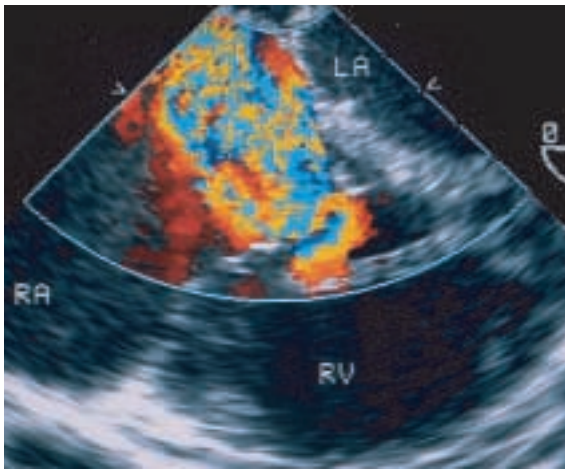

Figure 3

Pre-operative transoesophageal colour Doppler echocardiography, showing severe tricuspid valve regurgitation.

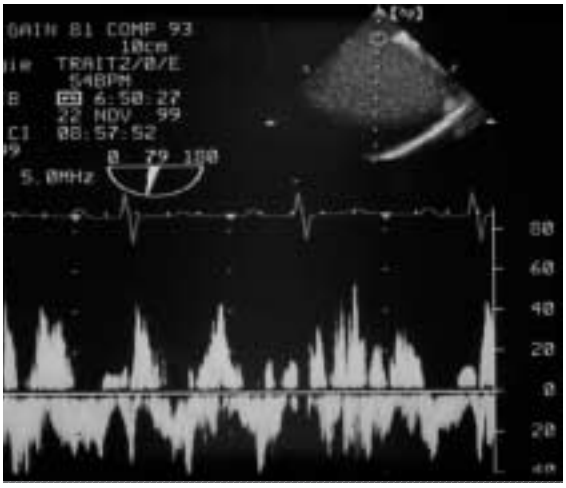

Figure 4

Pre-operative transoesophageal colour Doppler echocardiography, showing bi-directional (right-to-left and left-to-right) shunt trough the atrial septal defect.

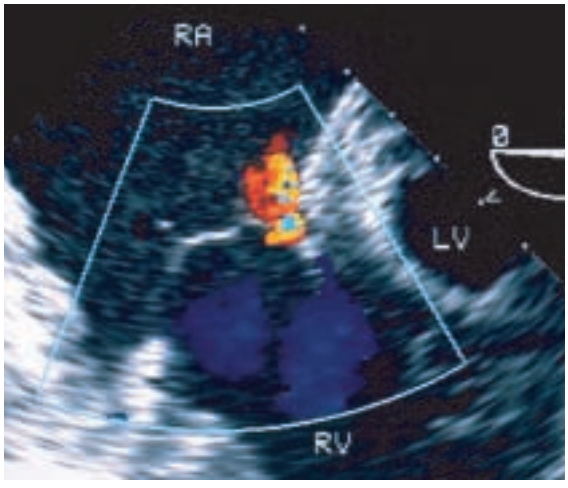

Figure 5

Post-operative transoesophageal colour Doppler echocardiography, showing reduced degree of the tricuspid valve regurgitation.

Table 1
Echocardiography parameters (mean data).

	RV diameter mm/m² BSA	RA volume cm³	tricuspid valve regurgitation	LV diameter mm/m² BSA	LV E.F.	LV S.F.
pre-operative	36.5	387	4/4	26.3	58%	25%
post-operative	22.7	116	2/4	29.8	77%	40%

BSA = body surface area; E.F. = ejection fraction; LV = left ventricle; RA = right atrium; RV = right ventricle; S.F. = shortening fraction

Results

There were no early or late deaths in a mean follow-up of 33 months. In all there was complete regression of the cyanosis due to relief of the intra-atrial right-to-left shunt and of the signs of heart failure. Postoperative echocardiography showed a reduced degree of tricuspid valve regurgitation (figure 5), substantial reduction of the size of right atrium and ventricle and improvement of the left ventricular function (table 1).

Discussion

In patients with Ebstein's anomaly referred late for surgery with severely compromised right ventricular function or even with reduced biventricular function, conventional biventricular repair is accompanied by heightened mortality and morbidity and poor functional results.

When a biventricular type of repair (systemic and pulmonary circulations in series, completely separated, each supported by a ventricle) is not suitable, the presence of a relatively hypoplastic and/or malfunctioning right ventricular chamber, inadequate to sustain the entire systemic venous return but capable of managing part of the systemic venous return, permits a one and a half ventricular repair.

The haemodynamic pattern of a one and a half ventricular repair is characterised by systemic and pulmonary circulations in series, completely separated, with systemic circulation fully supported by the left ventricle, pulmonary circulation dependent on a superior cavo-pulmonary connection (bi-directional Glenn) for the superior vena cava return and supported by a hypoplastic/dysfunctioning right ventricle for the inferior vena cava return. In order to perform a cavo-pulmonary connection, normal pulmonary artery pressure and resistance is indispensable.

Experimental background

Many years ago Starr [4] demonstrated in an experimental study that even a severely damaged right ventricle could positively contribute to the pulmonary circulation. Ilbawi [5] showed that the right ventricle even with a volume reduced to 30% of normal still had favourable haemodynamic effects on the pulmonary circulation. Recently Danton [6] evaluated the possibility of utilising a malfunctioning right ventricular chamber to manage only part of the venous return.

Clinical background

Despite the main goal of a congenital heart surgeon being to provide a biventricular repair whenever possible, the clinical use of a hypoplastic ventricular chamber to manage part of the systemic or venous return was first reported for both the left [7] and right [8] ventricular hypoplasia almost simultaneously.

Advantages

The main advantages of incorporating a hypoplastic (pulmonary) ventricle to partly support the pulmonary circulation are the following:
- ability to increase the cardiac output;
- adaptation to exercise;
- maintenance of pulsatile flow in the pulmonary circulation;
- flexibility to increased pulmonary vascular resistance;
- circulation at low venous pressure in the inferior vena cava system;
- ability of a hypoplastic right heart complex to adequately handle the reduced pre-load.

Indications

Following the initial clinical reports on the relative hypoplasia of both the left [7] and the right [8] ventricle, several other authors reported the application of one and a half ventricular repair in a huge variety of indications, mostly for problems related to the right ventricle, such as right ventricular morphology (small size) or dysfunction, acute as well as chronic [9–13].

The application of one and a half ventricular repair in Ebstein's anomaly deserves special comment. In this malformation, thanks to the volume unloading of the dysfunctioning right ventricle, the addition to a conventional repair of a superior cavo-pulmonary connection (bi-directional Glenn), allows the hypoplastic or dysfunctioning right heart complex to adequately handle the re-

duced pre-load. Furthermore, by reducing the systemic venous return by a third, a more aggressive surgical repair of the tricuspid valve without the risk of iatrogenic tricuspid stenosis is feasible, as confirmed by our experience.

These observations have been made independently of the underlying anatomy or the adopted surgical technique, but have been based upon the very poor right ventricular function [14].

Long term results

The major concern in the extended application of one and a half ventricular repair is the currently limited knowledge of the long term results, due to the relatively recently utilisation of this surgical approach and to the reduced number of clinical reports in this regard [15].

International Registry

In order to collect as much information as possible on potential indications, early and long term results, we have created an "International Registry for One And a Half Ventricular Repair". The Registry was officially announced at the last World Congress of Paediatric Cardiology and Cardiac Surgery in Toronto in 2001, and announcements appeared in other journals (Cardiol Young 2001; 11:582. Asian Cardiovasc Thorac Annals 2001; 3).

Information on the Registry is available at the following e-mail address: oneandhalf@chuv.hospvd.ch

Only data collection and analysis of a large number of patients will allow better clarification of the possibilities offered by one and a half ventricular repair and therefore facilitate future decision making progress with regard to this promising surgical approach, including patients with Ebstein's anomaly.

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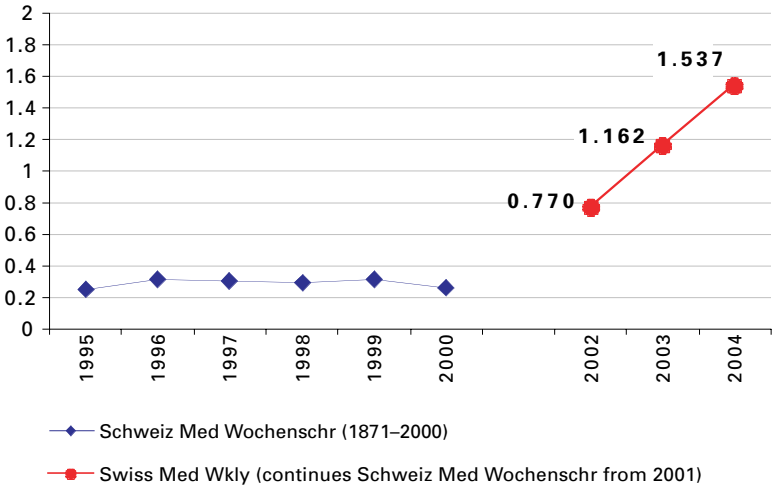

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