INTRODUCTION

There is a continuing need to perform inflow correction of “simple” TGA in a large subset of patients, who are not candidates for the arterial switch operation, particularly in developing countries. This is because of a lack of resources, centres and expertise to perform the neonatal switch operation. This subsequently results in the “late” presentation of patients who are severely cyanosed and have thin-walled, banana-shaped left ventricles which are not suitable for switching.

Inflow correction by the Mustard1 and Senning2 operations can transform the lives of these patients. However, there is continuing concern about the relatively impaired exercise capacity and long-term results of these patients, thought to be due to the fact that the right ventricle is not suitable for support of the systemic circulation. There is growing evidence that the right ventricle can function normally into old age in certain patients with corrected TGA (AV and VA discordance). Ourselves and others3-5 believe that the less-than-optimal results after the Mustard operation are, at least in part, due to an impaired pattern of left and right ventricular filling, produced by the relatively narrow, rigid, non-contractile channels in the atria, resulting in significant loss of the conduit, reservoir and contractile functions of the atria.

In an attempt to correct this, we have modified the Mustard operation to include a larger part of the contractile atrial wall into the SVC and IVC channels including both atrial appendages. We have previously described the three dimensional shape and function of the atria and ventricles following the unmodified Mustard operation6.

PATIENT AND METHODS

A three-year-old female patient presented with a history of cyanosis since birth. She was diagnosed as D-TGA with ASD and small PDA. She had balloon atrial septostomy on the fifteenth day of life. On presentation the oxygen saturation was 61%. Echo showed transposition of the great arteries with the aorta anterior and to the right, good size inter-atrial communication and deconditioned left ventricle.

A Mustard (atrial switch) operation was performed where the superior and inferior vena cavae were directed to the mitral valve using a trouser-shaped patch of autologous pericardium, while the pulmonary veins drained to the tricuspid valve. The atrial appendages were kept in the systemic venous channel, while the coronary sinus was kept in the pulmonary venous channel.

The patient had a smooth postoperative course and was discharged from the hospital one week after the operation, her pre-discharge echo showed good ventricular function with no systemic or pulmonary venous obstruction.
IMAGING AND 3D MODELLING

The size and shape of the intra-atrial channels and of both ventricles during the different parts of the cardiac cycle play an important part in the Mustard operation. Atrial function, pattern of atrioventricular flow and ventricular filling were evaluated using postoperative computed tomography (CT) angiography and echocardiography.

The ventricular end-systolic and end-diastolic phase were identified using ECG gating and CT-derived volumes for the four chambers. 3D modelling of the right and left side was used to visualize the supra-mitral and supra-tricuspid chambers. Images stored in DICOM format were imported into a commercial medical image processing tool Mimics Research version 17.0 (Materialise, Leuven, Belgium) for semi-automated segmentation and reconstruction of the blood volume. Further processing allowed isolation of the four heart chambers with boundaries defined at the valve levels. Those models were exported in 3-matic 9.0 (Materialise, Leuven, Belgium) to trim the inferior vena cava, superior vena cava and pulmonary veins. The final volumes could be measured or re-exported into Mimics for visualization. The same processing was achieved for all 10 phases across the heart cycle. By rotating the model and observing the dynamic changes, particularly of the left and right atrial appendages as well as the intra-atrial channels, the influence of the New Mustard operation is visualized. Echocardiographic images were acquired to evaluate the pattern of filling in the intra atrial channels.

RESULTS

This study serves to document for the first time the three-dimensional structure of the neo-atrial components after the modified Mustard operation and discuss their functional significance.

The neo-systemic venous (supra-mitral) chamber

This comprises an upper and lower component (Fig. 1), which represent the modified superior and inferior vena cava channels respectively. The upper component shows preserved SVC/right atrial junction designed to protect the SA node and its arterial supply. The upper component incorporates the right and left atrial appendages which provide reservoir and contractile function to the supra-mitral chamber.

The lower component of the systemic venous atrial chamber comprises the entry of the IVC and an adjoining “pouch” of atrial cavity designed to further increase the reservoir and contractile function of the supra-mitral chamber (Fig. 1). Blood from the upper and lower chambers passes smoothly to a relatively small collecting chamber, on to the mitral orifice, without the streams “colliding”.

Figure 1. 3D model of the neo-systemic venous (supra-mitral) chamber with an upper component incorporating the right and left atrial appendages (top, left: during ventricular end-systole at 40% of the cardiac cycle; bottom, left and right: during ventricular end-diastole at 90%).
The neo-pulmonary venous (supra-tricuspid) chamber
The four pulmonary veins open in a relatively large posterior chamber (Fig. 2), which communicates freely with an equally large anterior chamber which lacks an appendage. The combined anterior and posterior chambers provide reservoir and contractile functions, the blood passes directly into the tricuspid valve when it is open.

Figure 2. 3D model of The neo-pulmonary venous (supra-tricuspid) chamber with four pulmonary veins open in a relatively large posterior chamber communicating freely with an equally large anterior chamber which lacks an appendage (top, left: during ventricular end-systole; bottom, left and right: during ventricular end-diastole).

Pattern of filling of the left ventricle
Colour flow mapping of the left ventricular inflow at the level of the mitral valve, showed unimpeded laminar flow (Fig. 3). Importantly, pulsed Doppler interrogation of flow across the mitral valve showed a clear wave, indicating neo-atrial contribution to ventricular filling (Fig. 4).

Figure 3. Unimpeded laminar flow at the left ventricular inflow at the level of the mitral valve.
Right and left ventricular shape and function

Three-dimensional determination of size, shape and function (Fig. 5) showed good diastolic volume of both ventricles with no evidence of “banana”-shaped left ventricle. Both ventricles contracted very well as indicated by the small systolic volumes.

Limitations of this study include the fact that all data are derived from one patient, and that detailed hemodynamic measurements, electro-anatomical mapping and fluid dynamics were not studied. These limitations need to be addressed in future studies.

CONCLUSIONS AND FUTURE STUDIES

This investigation strongly suggests that the modified Mustard operation produces superior flow dynamics due to enhancement of conduit, reservoir and contractile functions of the neo atria, which should translate into enhanced survival and quality of life. Long-term results with a larger cohort of patients is required to validate these concepts.

REFERENCES


