INTRODUCTION

Tetralogy of Fallot (TOF), also called blue-baby syndrome is one of the most common congenital heart defects in children after infancy and is estimated to account for 10% of all congenital heart defects [3]. TOF consists of four interrelated lesions: i) ventricular septal defect ii) Pulmonary stenosis iii) Right ventricular (RV) hypertrophy and (iv) Overriding Aorta [3]. TOF has been successfully repaired for several decades (Fig. 1). There are now an estimated 100,000 adult “repaired TOF” patients in the United States alone. As a result, long-term sequelae of the disease and repair have become important clinical issue. Specifically, residual pulmonary valve insufficiency (PI) is one such accepted and often unavoidable sequela. PI, when severe, abnormally alters the RV loading conditions, thereby triggering RV hypertrophy and dilatation. In turn, RV dilatation can evolve into irreversible RV myocardial contractile dysfunction, and has been related to sudden death in many “repaired TOF” patients. To normalize RV loading conditions, pulmonary valve replacement is often necessary and should be performed prior to the onset of irreversible RV myocardial damage.

Whitehead et. al [1] have employed control volume analysis to study energy losses in fontan patients and reported marked difference in energy losses between normal and fontan patients. The goal of this study is to evaluate the pulmonary circulation in TOF patients in adulthood by calculating the work done by the RV and the total power output for the normal and the repaired TOF patient.

METHOD

Phase encoded MRI data were obtained for patients with tetralogy and one without tetralogy problem. Fig. 2 compares MRI images of a normal patient and a repaired TOF with severe PI and severe RV dilatation. For this study we compare the RV pressure-volume (P-V) work for a normal and a TOF patient. The complexity of the RV loading conditions in TOF patients is expected to be more complex due to the presence of residual PI and RV dilatation.
in our study lies with obtaining the catheter data for the subjects. From the phase encoded MRI data, contours for RV interior are traced out along sequentially stacked cross-section locations of the heart and at different time instants to generate RV volume versus time data. The pressure-time and volume-time data are synchronized using EKG by identifying the beginning of the systole and is used to construct the P-V diagram. The work done by the right ventricle is taken as the area enclosed by the P-V diagram. This work is taken as the energy that the RV imparts to the blood that is being pushed into the PA. Likewise, main pulmonary artery (MPA) flow and pressure versus time data is obtained from additional MRI and catheter data at MPA location. The total power in the blood is computed using transient pressure and flow data and by applying Bernoulli’s equation [1,4].

RESULTS AND DISCUSSION

The P-V work done by the RV for a normal and a repaired TOF patient is tabulated in Table 1 which are computed from Fig. 3. The column 4 gives the absolute values of P-V work done by the RV in energy units and the column 5 scales the work done by body surface area of each patient (BSA) to compare between patients with different physical characteristics [4].

Data in Table 1 shows that the work done by RV is almost 30% less for a repaired TOF patient compared to a normal patient with similar stroke volume and BSA. This shows that the RV of a repaired TOF patient is able to do less work to pump blood inspite of elevated cardiac pressure. It is also noted from the P-V plot (Fig.3), that the RV of a repaired TOF patient operates at a higher peak systolic pressure (30mm Hg) than a normal subject (25mm Hg). The isovolumetric contraction of the RV of a tetralogy patient is much less pronounced compared to normal subject. This is attributed to the back circulation, which could be a consequence of either the absence, or the deteriorated state of the PV, which results in blood flowing back into the RV and hence elevated pressure levels in RV during the diastolic relaxation phase. This results in a narrower PV loop and thereby much less energy is imparted to the blood flowing out of the RV into the PA. The total MPA power output plot (Fig. 4) also support the above conclusion showing significant negative power in the case of repaired TOF patient. Also, our study shows that the MPA flow and pressure are out of phase in repaired TOF patients compared to a normal patient (Fig. 5), further illustrating the loss of energy in PA.

In future, the fluid structure interaction (FSI) study of this problem may reveal more characteristics of flow in compliant PA.

REFERENCES


