

SHOULD ALL PATIENTS WITH CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES (CC TGA) UNDERGO ANATOMIC REPAIR?

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Background: Congenitally corrected transposition of the great arteries (cc TGA) is characterized by the combination of atrio-ventricular and ventriculo-arterial discordance. The morphologic left ventricle supports the pulmonary circulation, while the morphologic right ventricle supports the systemic circulation. Long term follow up has demonstrated diminished survival associated with a failing systemic morphologic right ventricle and or associated systemic atrioventricular valve. (1-4).

Surgical approaches for cc-TGA include two main strategies: physiologic and anatomic repair. A physiologic repair is defined by managing the associated VSD and or pulmonary stenosis while leaving the right ventricle in the systemic circulation. An anatomic repair restores the morphologic LV to the systemic circulation either by an arterial and atrial switch or via a Rastelli and atrial switch. Failure of the systemic ventricle (the morphologic RV) in congenitally corrected transposition of the great arteries (ccTGA) is problematic in those not requiring an operation and in those having a physiologic repair. A contemporary change in strategy in our institution allows for a long term and more recent analysis of outcomes.

Objective: To test the hypothesis that patients undergoing an anatomic repair for ccTGA will have superior survival and systemic ventricular function.

Method: Between 1982 and 2015 (34 years), 165 patients with biventricular ccTGA were studied. Groups were physiologic repair (PR, n=89), anatomic repair (AR, n=38), and no intra cardiac repair (NICR, n=38). To gain perspective on Fontan outcomes, left hand topology Fontan patients were reviewed (n=35).

Result: Median age at "definitive repair was 5 years (2-13) for physiologic repair, 3 years (1-8) for anatomic repair, and 4 years (3-6) for the Fontans (p=0.04). Median follow up in years was: 13.5 (6.5-23.3) PR, 3.4 (0.7-9.7) AR, 7.5 (1.7-15.7) Fontan, and 11.8 (4.9-20.9) NICR.

Survival: For the entire cohort, 20 year freedom mortality or transplant was 65% (PR), 53% (AR), and 73% for Fontan. (See figure 1). In the most recent era (2000-2015), 10 year freedom from mortality or transplant was 86.5% (PR), 79% (AR) and 100% for Fontan, P=0.17. Survival in the no intracardiac repair group was 94.55 at 25 years. There were 10 cardiac transplants performed for failure of strategy. Four each for PR (4.4%) and AR (10.5%). There were 2 for the no intra-cardiac repair group (5.2%).

Subgroup analysis showed no difference in transplant free survival between arterial switch/mustard and the Rastelli/Mustard (p=0.63).

Patients in the staged procedure group are those who underwent any palliative procedure and are awaiting definitive surgery or are deceased prior to definitive surgery. The hearts are biventricular in 19/34 (mortality = 3, 15.8%). The remaining 15/34 are single ventricle pathway patients. There were 9 pre definitive stage deaths (60%).

Function of the systemic ventricle: Systemic ventricular function pre definitive operation, post definitive procedure, and at last echocardiographic study are shown in figure 2. The following percentage of moderate to severe reduction are noted by groups: 38.7% (PR), 28.1% (AR), 23.4% (NICR), and 8.3% Fontan. For those not having intracardiac repair it is their first and last echocardiogram.

All groups showed some change in ventricular function over time. Echocardiographic data is more reflective of procedures subsequent to the year 2000.

Systemic atrioventricular valve function pre definitive operation, post definitive procedure, and at last echocardiographic study are shown in figure 3. All groups had some degree of moderate to severe regurgitation of the systemic atrioventricular valve prior to the definitive operation: 16.6% in (PR), 17.1% in (AR), 5% in Fontan, and 12.9% in (NICR). Immediately following the definitive operation there was improvement of systemic AV valve regurgitation except in the physiologic repair group.

At the last follow up there was valve function deterioration with the following percentages showing moderate to severe systemic AV valve regurgitation: (53.8% (PR), 28.1% (AR), 34.3% (NICR) and 8.3 Fontan.

Independent predictors: There were no independent predictors of mortality when comparing the entire cohort of AR, PR and the Fontan groups. Independent markers of systemic ventricular function were year at operation >2000 (OR 0.14, p=0.01) and early post repair depressed systemic ventricular function (OR 12.4, P 0.03).

Independent predictors of systemic atrioventricular valve dysfunction were Fontan procedure (OR 0.12, p= 0.02) and early postoperative atrioventricular valve dysfunction (OR 9.0, p= 0.01).

Conclusion: In this cohort, anatomic repair did not result in superior survival or superior systemic ventricular function. All groups showed a decline in systemic ventricular function and an increase in systemic atrioventricular valve regurgitation over time. Given greater than 90% 25 year survival in the no intra cardiac repair patients, prophylactic double switch surgery for isolated ccTGA should be viewed with caution.

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Figure 1 transplant free survival of the entire cohort

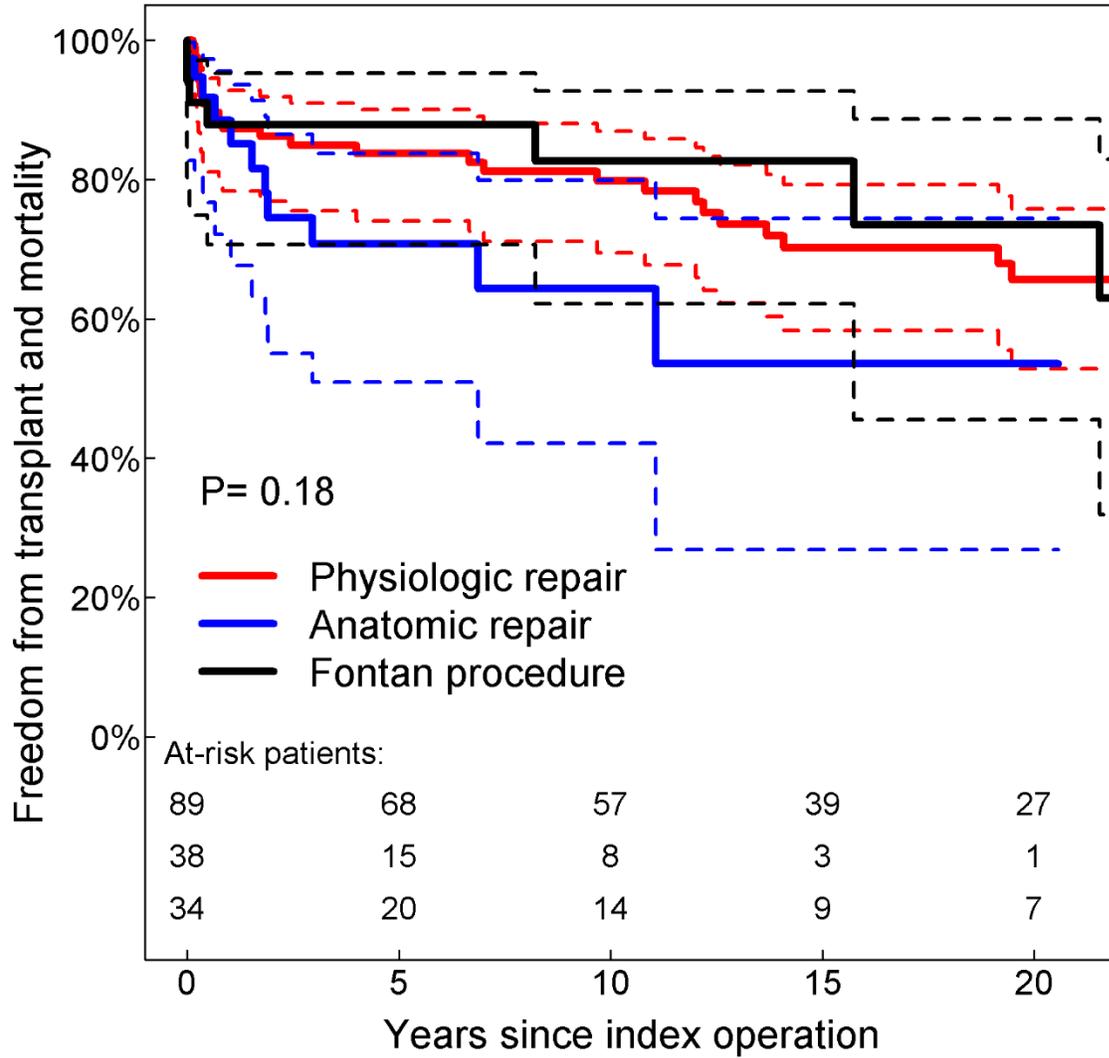


Figure 2 Echo data of the systemic ventricular function for the entire cohort.

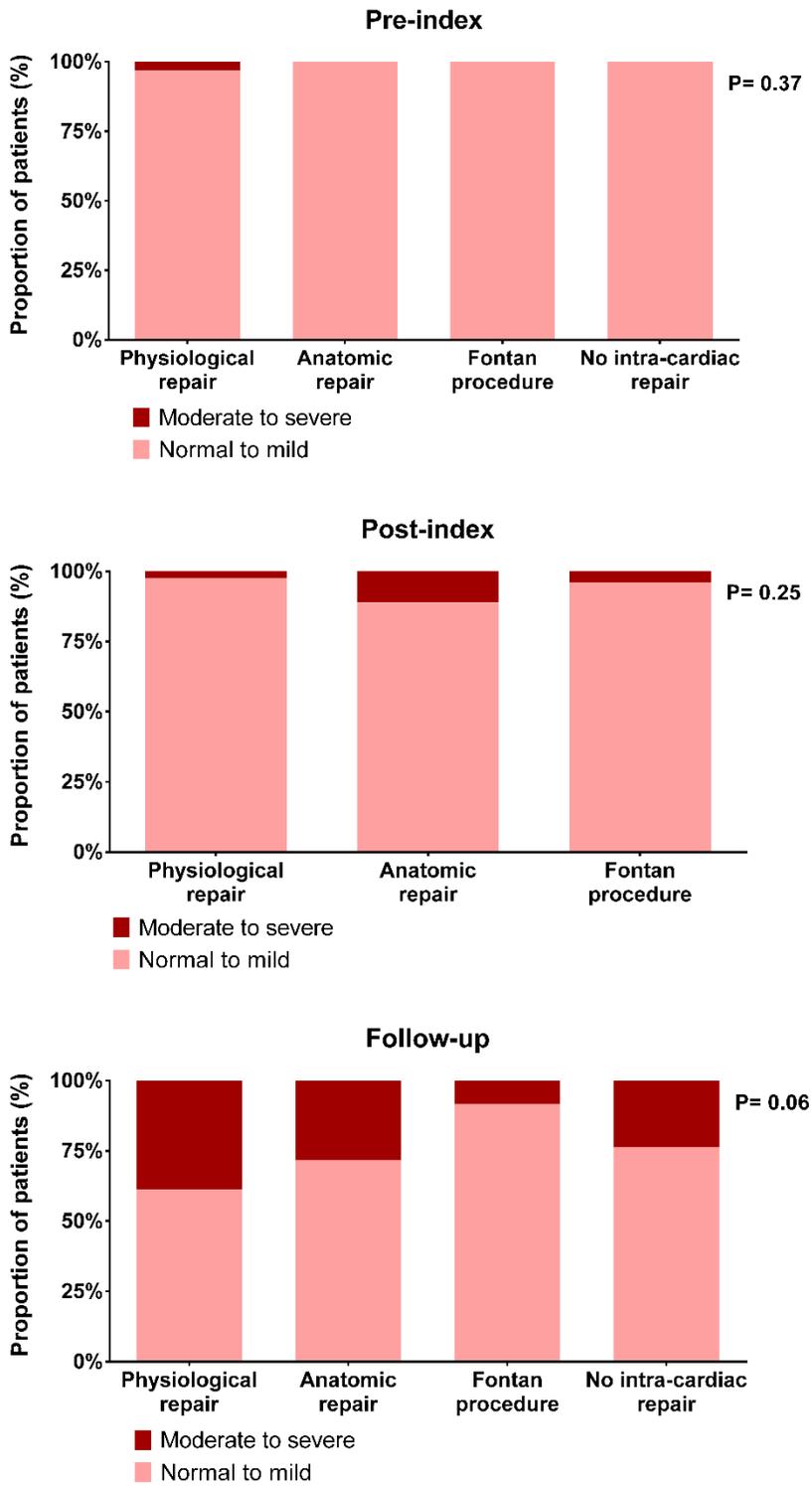


Figure 3 Echo data of the systemic AV valve function of the entire cohort

