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Heart Institute

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Heart Transplant vs Complex Repair in the High Risk ACHD Patient

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ACHD Symposium, Queenstown, New Zealand

Nov 30-Dec 2, 2017



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School of Medicine
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ONLY ONE DISCLOSURE



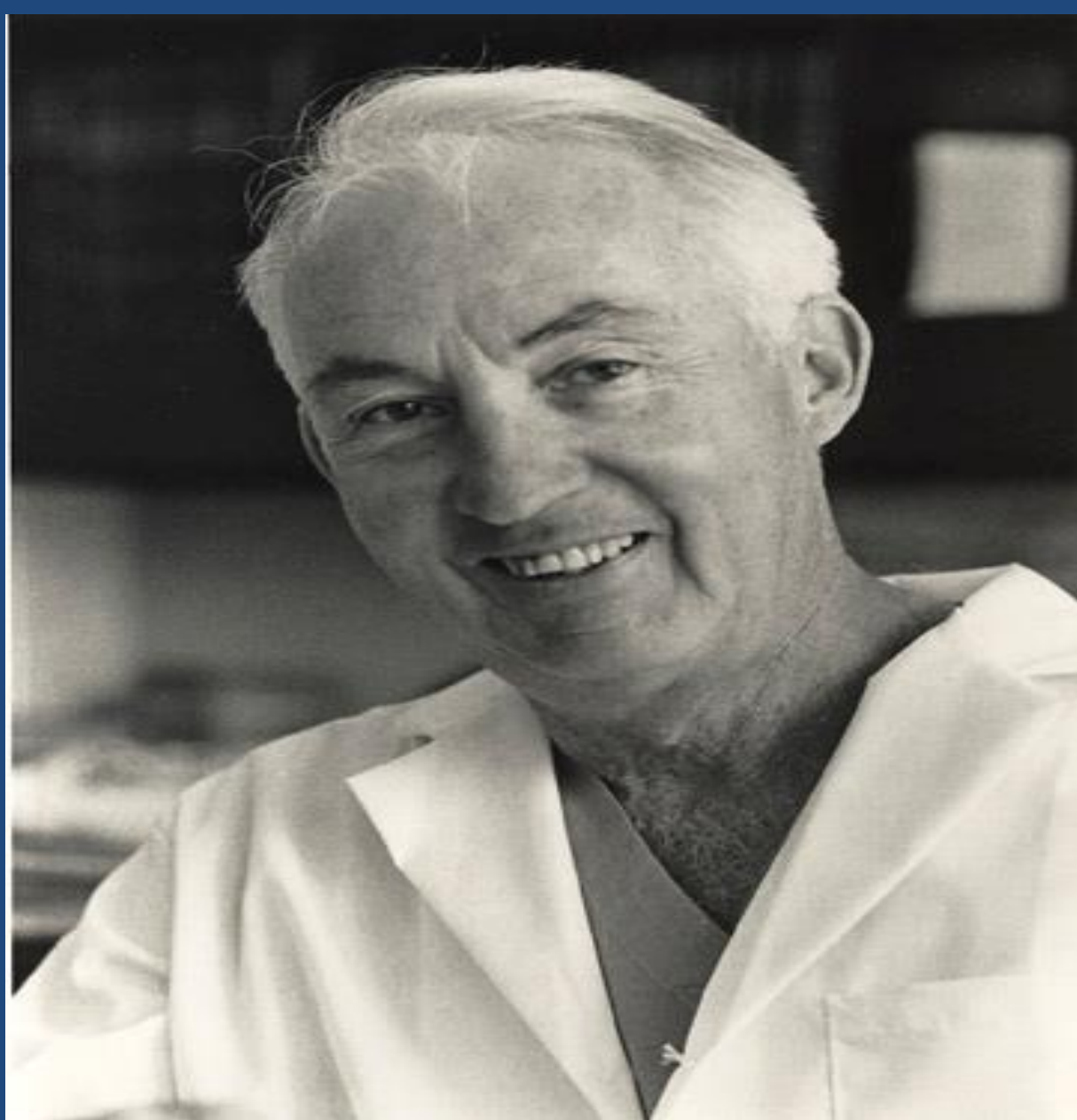
LET'S MOVE ALONG. THOSE ARE
SURGEONS WHO JUST GOT HERE
AND FOUND OUT THEY'RE NOT GOD.

NEW
ARRIVALS





Thomas Earl Starzl MD, PhD



Norman Shumway MD



Leonard L. Bailey M.D.

FATHER OF PEDIATRIC HEART TRANSPLANT

Heart transplant or high risk complex surgery- which is better?

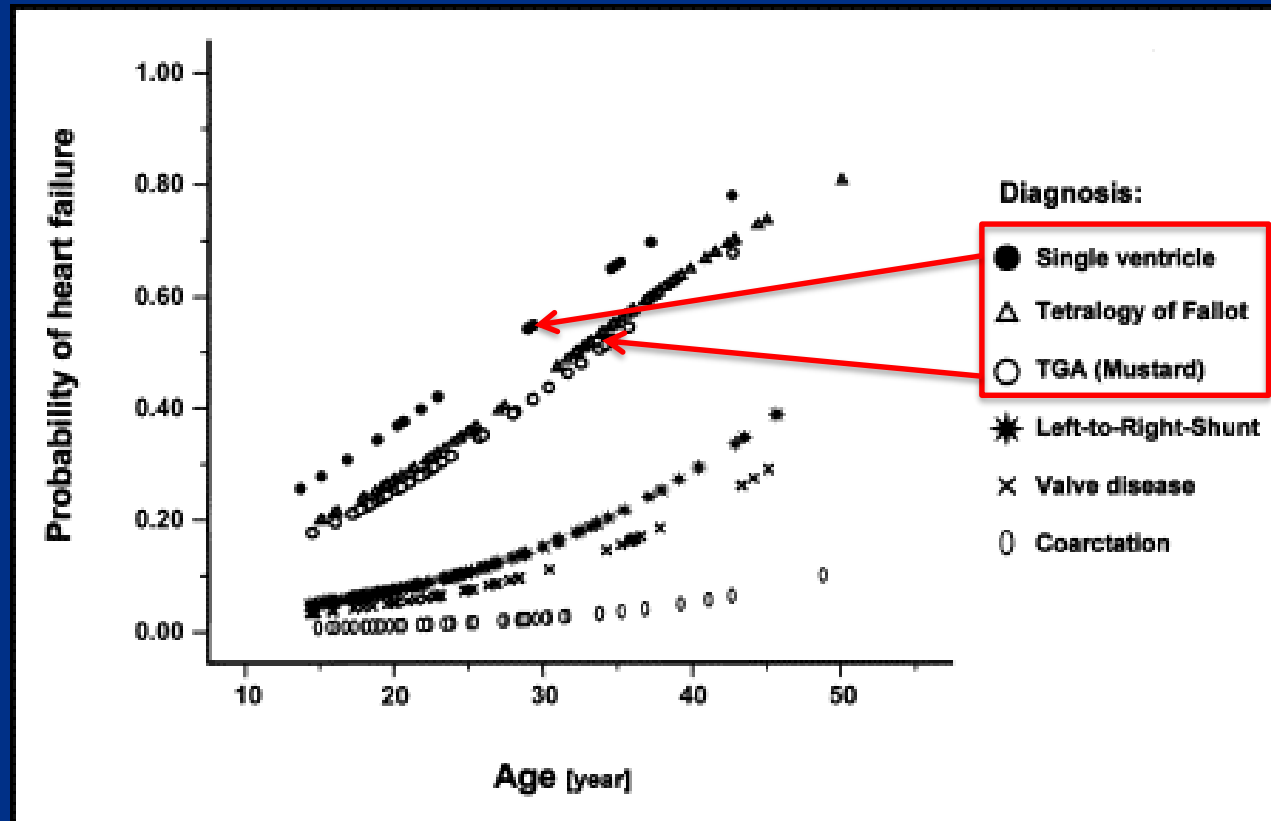


The Question

The Answer

WHICH IS SAFEST FOR
THE INDIVIDUAL
PATIENT?

Incidence of heart failure in adults with CHD after cardiac surgery



[Norozi et al. Am J Cardiol 2006]

Factors contributing to Long term cardiac dysfunction in ACHD

- Prolonged Volume overload
 - Intra and extra cardiac shunts , valvular insufficiency
- Prolonged Pressure overload
 - pulmonary and aortic stenosis and PHTN
- History of myocardial damage from previous surgery/surgeries
- Prolonged cyanosis
- Arrhythmias
- Effects of Aging especially on Single Ventricle:
“failing” Fontan, PLE, Plastic bronchitis, hepatic fibrosis or cirrhosis



Be careful of bad advice

High Risk Cardiac Surgery as an Alternative to Transplant or Mechanical Support in Patients with End Stage Heart Failure

Kawajiri et al. Toronto JTCVS 2017

- Study looked at subset of patients with LVEF < 20% and VO₂max < 14ml/min/m², initially referred for transplantation or ventricular assist device. Of 133 patients who then underwent high risk conventional cardiac surgery, 68 were transplant eligible, 65 were not.
- These patients were compared to 2nd group who underwent HTx or BTT-LVAD or 3rd group who had LVAD as destination therapy

E-mail: CLOSETBHAMPER@AOL.COM (and I'll send you a photo of my rat!)



“They said I wouldn’t survive the surgery to implant a regular pacemaker.”

High risk cardiac surgery in end stage HF

- Survival in the HTx-eligible group was equal to HTx-BTT/LVAD group
- Survival in the HTx-ineligible group was equal to the Destination Therapy group
- Mortality was 12% overall
- Patients who had HF on basis of ISCHEMIA did best. Coronary artery bypass patients had lowest mortality = 2.3%
- These patients are very different than ACHD patients who have complex structural issues (difficult anatomy, multi-operations, cardiac shunts, AVM's)

Is Alternative Cardiac Surgery an Option in Adults with Congenital Heart Disease referred for Thoracic Organ Transplant?

Harper et al, Newcastle upon Tyne, UK, EJCTS 43(2013) 344-351

- 126 ACHD patients assessed for thoracic organ transplant candidacy over 11 years
- 14(11%) underwent non-transplant cardiac surgery
- 4 groups: left sided lesions(4), right sided lesions(3), systemic right ventricle(5), single ventricle (2) both underwent Fontan Conversion

Is Alternative cardiac surgery an option?

- Results: 2 early deaths, 3 late deaths = 1year mortality of 28%
- Both single ventricle Fontan conversions died
- 3 of the 5 patients with systemic right ventricle died(all had VAD placed at time of surgery)
- Of the 9 patients alive: 3 have Heartware VAD's in place (from time of surgery)
- Therefore, only 6 of the 14(43%) had successful outcome with surgery alone

**GOOD ADVICE IS
SOMETHING A MAN
GIVES WHEN HE IS
TOO OLD TO SET A
BAD EXAMPLE**

FRANCOIS DE LA ROCHEFOUCAULD

Cite this article as: Poh CL, Cochrane A, Galati JC, Bullock A, Celermajer DS, Gentles T et al. Ten-year outcomes of Fontan conversion in Australia and New Zealand demonstrate the superiority of a strategy of early conversion. *Eur J Cardiothorac Surg* 2016;49:530–5.

Ten-year outcomes of Fontan conversion in Australia and New Zealand demonstrate the superiority of a strategy of early conversion[†]

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Received 5 October 2014; received in revised form 4 February 2015; accepted 19 February 2015

Abstract

OBJECTIVE: To investigate the benefits of a strategy of early Fontan conversion.

METHODS: Using the Australia and New Zealand Fontan Registry, retrospective analysis of their long-term follow-up data was performed.

RESULTS: Between 1990 and 2014, a total of 39 patients underwent surgical conversion in 6 centres at a median age of 23.8 years (IQR: 19.3–28.2), 18.7 ± 5.0 years post-Fontan. One centre tended to perform conversion earlier: interval since first documented arrhythmia 2.9 ± 4.0 vs 4.0 ± 4.2 years, average NYHA Class 2 ± 0.4 vs 3 ± 0.9 ($P = 0.008$), mean number of preop anti-arrhythmics 1 ± 0.4 vs 2 ± 0.6 ($P = 0.05$). Two patients underwent conversion to an extracardiac conduit only, while 36 had concomitant right atrial cryoablation, of which 16 also had pacemaker implantation. Nine patients suffered major cardiac-related complications (7 low output syndrome, 3 ECMO, 3 acute renal failure, one stroke) (2/17 from the early conversion centre and 7/22 of the others; $P = 0.14$). Four patients died in hospital (10.3%) and 4 late after a median of 0.9 years [95% confidence interval (CI): 0.5–1] after conversion. An additional 2 patients needed transplantation at 1 and 8.8 years after conversion, respectively. The 10-year freedom from heart transplantation was 86% (95% CI: 51–97%). Outcomes from the centre with an early conversion strategy were significantly better: 8-year freedom from death or heart transplantation was 86% (95% CI: 53–96) vs 51% (95% CI: 22–74; log-rank $P = 0.007$). Eight additional patients required pacemaker implantation and 5 had arrhythmia recurrence.

CONCLUSIONS: Fontan conversion is associated with lasting survival outcomes up to 10 years after conversion. A strategy of surgical conversion at earlier stage of failure may be associated with better survival free from transplantation.

Keywords: Fontan conversion • Atriopulmonary Fontan • Long-term outcomes • Fontan procedure

INTRODUCTION

the ideal surgical palliation, making up less than 1% of Fontan palliation in Europe and North America [3]. The population of

THE PEOPLE WITH THE
BEST ADVICE
ARE USUALLY THE ONES
WHO HAVE BEEN
THROUGH THE MOST

Evidence of ventricular dysfunction or “failed Fontan physiology” is a contraindication for Fontan Conversion, and I would recommend Heart Transplantation.



MORE
Good Advice



High Risk ACHD

- Prevalence of severe ACHD is increasing due to improvements in surgical repair and particularly specialized ICU care
- But, this is Palliative! Especially for Single Ventricle and systemic RV patients
- Heart Failure is the leading cause of death in ACHD: 26%-42%
Sudden death 2nd at 19%
- Heart Transplantation is increasingly used for end stage ACHD patients representing 1.8% of recipients worldwide in 1992-2000, but 3.1% of recipients in 2006-2013

VERY HETEROGENEOUS GROUP



Referrals for Heart Transplantation in Adult Congenital Heart Disease

- High risk operation required to repair or palliate
- Eisenmenger Syndrome(heart-lung)
- Tetralogy of Fallot(repaired or palliated) and other cyanotic complex Congenital Heart Disease
- Univentricular heart with or without Fontan circulation
- Systemic right ventricle i.e, CCTGA or TGA repaired with Atrial(Senning or Mustard) Switch

Facts about Heart Transplantation in Adult Congenital Heart Disease

- 10-20% of Adults with Complex CHD will ultimately require heart or heart-lung transplant
- Compared to adults with acquired heart disease, those with ACHD face increased perioperative risk 18% vs 7% mortality
- ACHD patients waiting for transplant are more likely to be assigned a lower wait list status
- ACHD patients less likely to receive AICD's and face higher risk of sudden death on wait list

- ACHD patients are less likely to be dependent on inotropes or mechanical cardiac support at the time of listing. Less MCS use may be related to higher difficulty due to previous operations, difficult anatomy
- Patients are younger, have less comorbidity
- ACHD patients have higher PRA's (class II antigens > 10%)

Eligibility for Heart Transplantation in ACHD

- Peak oxygen consumption (Vo_2max) $< 14\text{ml/kg/min}$
- Predicted peak oxygen consumption $< 55\%$
- $\text{PVR} < 6$ Wood units / m^2 and transpulmonary gradient < 12 or positive response to modulation with vasodilator drugs i.e., O_2 , NO
- B type natriuretic peptide levels may be important, but not clear, since has no diagnostic value in systemic RV and normal in “Failed Fontan” with preserved ventricular function

Relative Contraindications for Heart Transplantation in ACHD Patients

- Obesity with pre-transplant BMI > 30 kg/m²
- Active smoking and/or substance abuse
- HIV positive
- Residual shunts resulting in high output failure post transplant i.e., aortopulmonary collaterals
- Severe peripheral vascular disease

Absolute Contraindications for Heart Transplantation in ACHD Patients

- Active Infections (active hepatitis C and viral replication)
- Severe metabolic disease
- Active malignancy
- Irreversible Multisystem Organ Failure
- Elevated PVR-not modulatory

Word of Caution

- Many ACHD patients particularly, post Fontan procedure, have poor exercise tolerance, and therefore results of exercise testing, including 6 minute walk, may be suboptimal.
- However, a blunted heart rate response to exercise(chronotropic incompetence) and attenuated rate of recovery of heart rate after exercise testing may carry an important prognostic value in these patients.

Another Word of Caution

It is difficult to accurately evaluate pulmonary vascular resistance in ACHD patients, especially single ventricle anatomy and post Fontan, with open fenestrations, low pulmonary blood flow, AVM's, aortopulmonary or veno-veno collaterals, and abnormal pulmonary vasculature

IT'S ALL PLUMBING!



Outcomes for Transplantation in ACHD

Congenital Heart Disease is a risk factor for increased morbidity and mortality in adult heart transplantation

- This is primarily due to the increased risk in the perioperative period from presence of previous surgical adhesions, aortopulmonary collateral vessels, increased PVR, complex anomalies of pulmonary and systemic venous return, and great artery malalignment.
- These factors lead to increased graft ischemia, perioperative bleeding, early graft failure

Outcomes in ACHD Patients Undergoing Heart Transplantation: a systematic review and meta-analysis

Doumouras et al, Toronto J Heart Lung Transplant 2016

- Analyzed 13 studies evaluating cause specific post transplant Mortality in ACHD patients vs non-CHD patients
- Results: death secondary to primary graft failure, stroke, and hemorrhage was significantly higher in the ACHD group
- In 3 studies that assessed Fontan/Glenn patients, 30 day mortality was significantly greater than in non-CHD patients 43.8% vs 14.4%

Morbidity following Cardiac Transplant for ACHD

UNOS DATA 41,849 adults 2.5%(1035) with ACHD

- CHD patients post transplant require reoperation for cardiac complications at higher rate than non-CHD patients
previous operation: 20.4% vs 13.7%
- CHD patients post transplant have higher incidence of post transplant dialysis
previous operation: 24.7% vs 10.8%

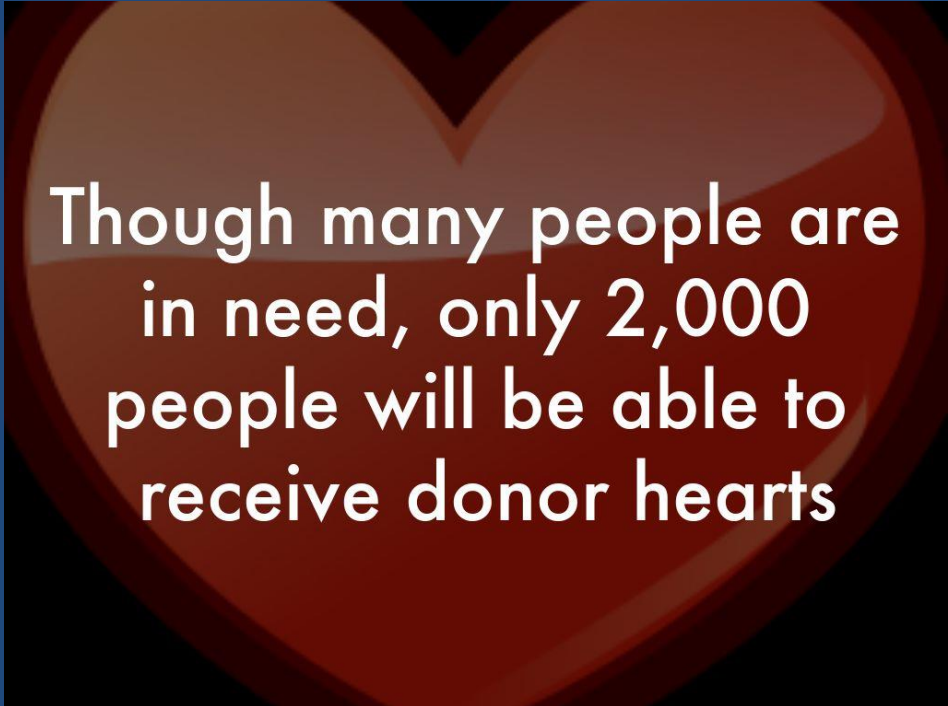
Davies et al: Listing and Transplanting Adults with Congenital Heart Disease (conclusions)

- MECHANICAL CIRCULATORY ASSISTANCE DID NOT IMPROVE WAIT LIST SURVIVAL IN ACHD PATIENTS
- ADULTS WITH CHD HAVE HIGH 30 DAY MORTALITY 17-19%

LIKELY DUE TO HIGHLY COMPLEX REOPERATIVE SURGERY AND OFTEN POOR PREOPERATIVE SYSTEMIC HEALTH

Multiple sternotomies, txp into single lung , pulmonary reconstruction, low serum albumin

With high early mortality and few donor hearts: Why transplant these patients with ACHD?



Though many people are in need, only 2,000 people will be able to receive donor hearts

Survival Paradox

Those ACHD patients who undergo a heart transplant and who survive 1 year have a better long term survival than non-congenital recipients

OUTCOMES after ORTHOTOPIC HEART and HEART-LUNG TRANSPLANTATION FOR CONGENITAL HEART DISEASE in Australia and New Zealand

Hatzistergos et al, JHLT April, 2013, vol 32, 4, S13-14

- 10 year survival for heart alone with CHD was 80%
- Median survival in the heart alone with CHD 19.7 years
- ISHLT median survival of 11 years for all heart transplant recipients overall

Classic Eligibility Criteria for Cardiac Transplant in the general Adult population

- End stage heart failure $EF < 25\%$, $VO_2\text{max} < 14\text{ml/min/m}^2$
- Life expectancy less than 2 years
- Failure of medical management
- No absolute contraindications

But should we use these for ACHD?



The Keys

To optimize outcome in ACHD

- Earlier consideration for heart transplant candidacy prior to end organ dysfunction, using nontraditional measures i.e., progressive cyanosis, exercise intolerance, and failure to thrive rather than predicted life expectancy
- Multidisciplinary approach and close collaboration between ACHD specialists, heart failure specialists, congenital and adult heart transplant surgeons, and medical specialists

Keys Continued

- Many ACHD patients have Body Mass Index < 18.5 at transplant. Optimizing nutritional and cardiac functional status is increasingly recognized as an important contributor for better outcomes.
- Again this speaks to earlier listing for heart transplant even though these patients don't fit classic criteria



Changing Face of End Stage Adult Congenital Heart Disease

- Considerations for Fontan conversion vs cardiac transplantation will disappear.
- Right Heart Failure in TGA Atrial Switch patients as an indicator for heart transplant will disappear.
- Patients with “failed Fontan” physiology will increase dramatically, requiring more complete evaluation of bystander organ dysfunction and raising the question of combined heart-kidney or heart-liver transplantation.

Who should have Combined Heart-Kidney Transplant?

- Combined Heart Kidney Transplant has increased dramatically in the last 2 decades with better results than heart alone with Renal Insufficiency.
- Renal insufficiency (creatinine clearance $< 40\text{ml/min}$) is a predictor for mortality in ACHD, both on Waiting list and post transplant.
- Exact criteria for HKT except for dialysis dependence are not well established, because difficult to predict recovery of renal function.
- Likely, best to consider early listing and combined HKT

Who Should have Combined Heart-Liver Transplant?

- Rare indication in ACHD at the present time
- As a guide , patients with evidence of cirrhosis who have normal synthetic liver function , normal hepatic venous anatomy, liver volume of >800 ml, only mild evidence of portal hypertension, and no hepatocellular carcinoma are candidates for heart only transplantation
- Combined heart-Liver transplant should be reserved for those patients who meet standard criteria for liver alone transplant

So What is the Answer to the Question?

- There are just not enough donor organs to treat all the patients who qualify for transplant and the waiting times and candidate numbers continue to increase.
- Despite a high perioperative risk, reparative procedures in Adults with Congenital Heart Disease may be an alternative option for highly selected patients(those who can not wait for Tx).
- Discussions concerning the role of MCS as destination therapy are already occurring and will become increasingly important for ACHD patients.



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TO AN OPERATING THEATRE NEAR YOU!



QUESTIONS ?