



**Stanford**  
MEDICINE

# Simultaneous Heart-Liver Transplant for the Treatment of Decompensated Fontan Patients

Seth Hollander, MD

Clinical Associate Professor, Pediatrics (Cardiology)

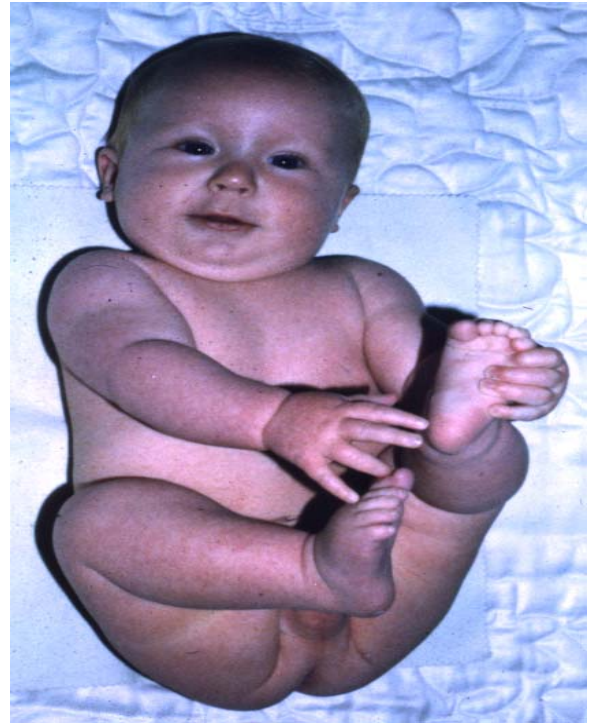
Medical Director, Cardiac Transplantation



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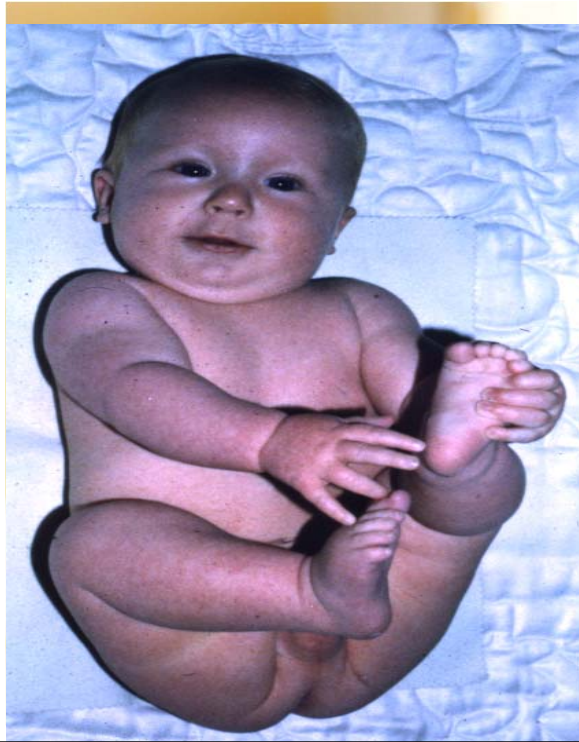




...it will



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## The Results of our “Success”



90% repaired CHD will survive into adulthood

Mechanisms heart failure are different than for non CHD adults.

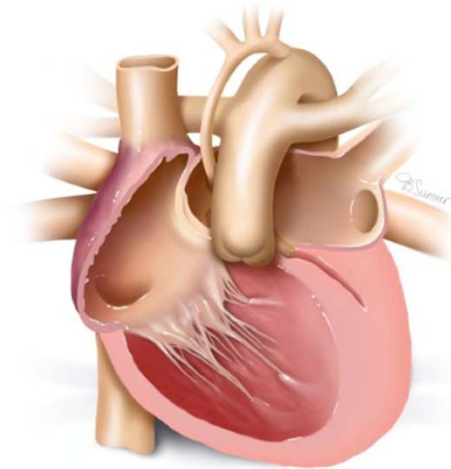
Medical therapy often fails...Increase in referrals for transplantation

3.3% of adult heart transplants (40% increase over previous decades)



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# Hypoplastic Left Heart Syndrome



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## Overview



What is the Fontan Operation?

Why does the liver "fail" in patients with Fontan circulation?

Heart-Liver Transplant Surgery

Outcomes

Immunologic Tolerance

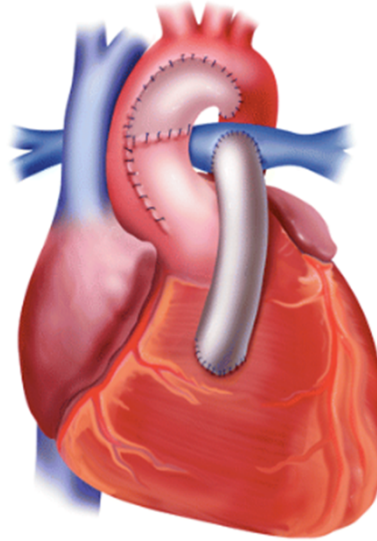
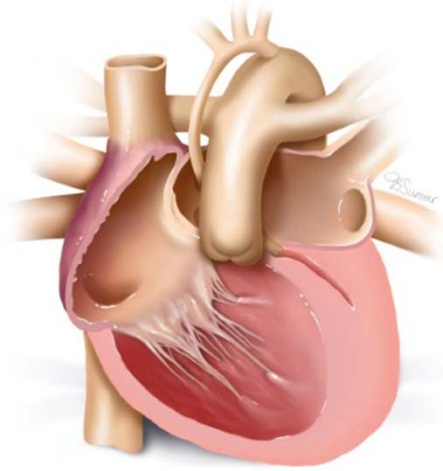
Ethics



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## The Fontan Operation for HLHS



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## Fontan Operation



Now standard of care for single ventricle disease.

Early mortality is low.

Survivors entering 20's and 30's.

Not a long-term solution.



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## History



Historically considered a contraindication to heart transplant secondary to concerns regarding bleeding, infection, varices.

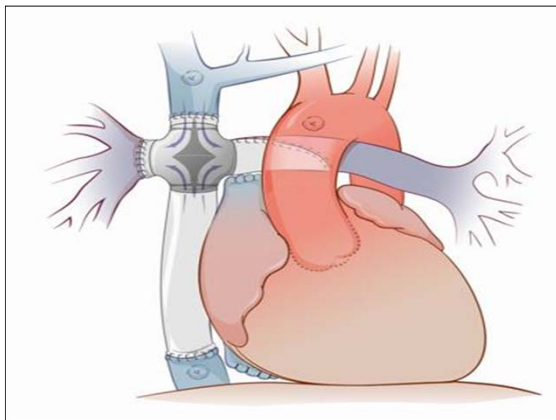
Liver failure in the setting of heart disease is increasing owing to improved long-term survival in patients with single ventricle disease.

Historical Contraindications = Current Indications



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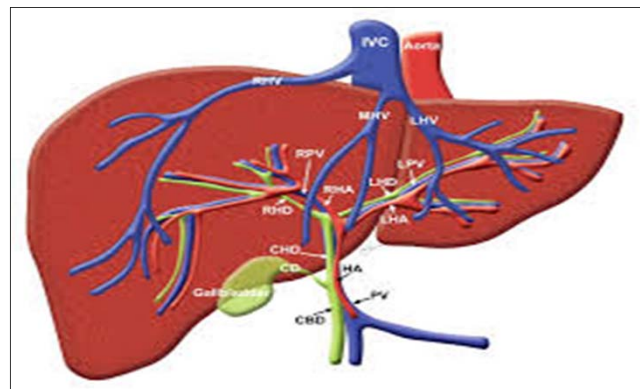
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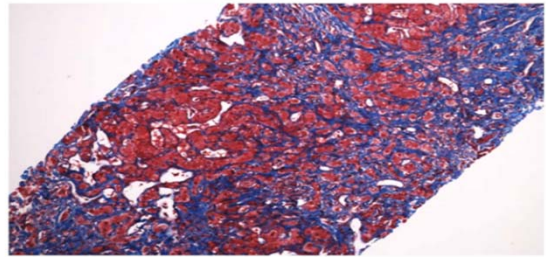
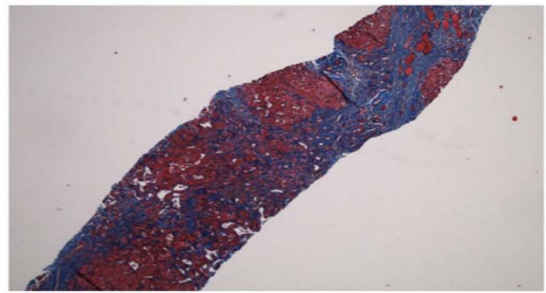
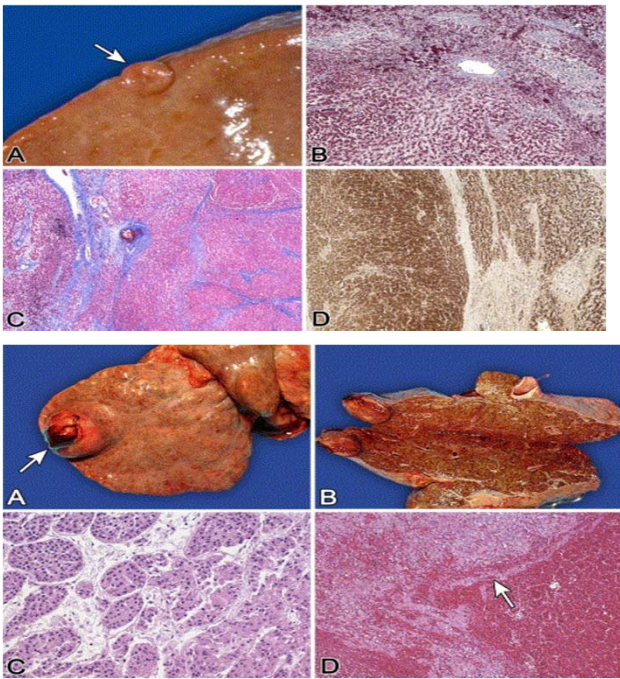
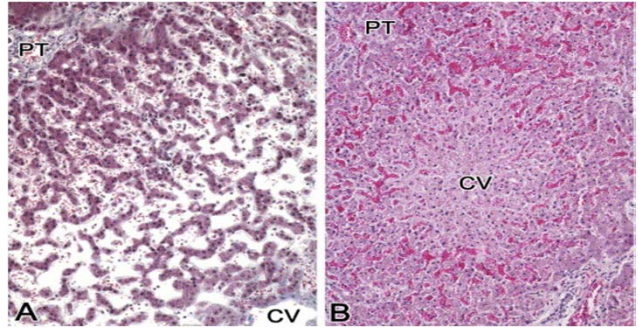
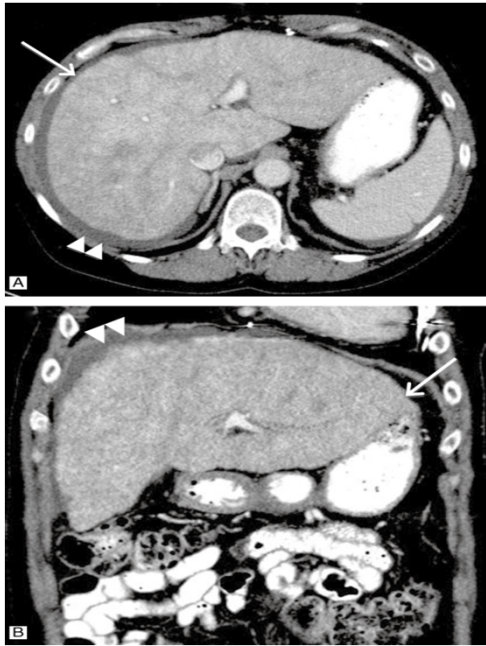


High Central  
Venous Pressure

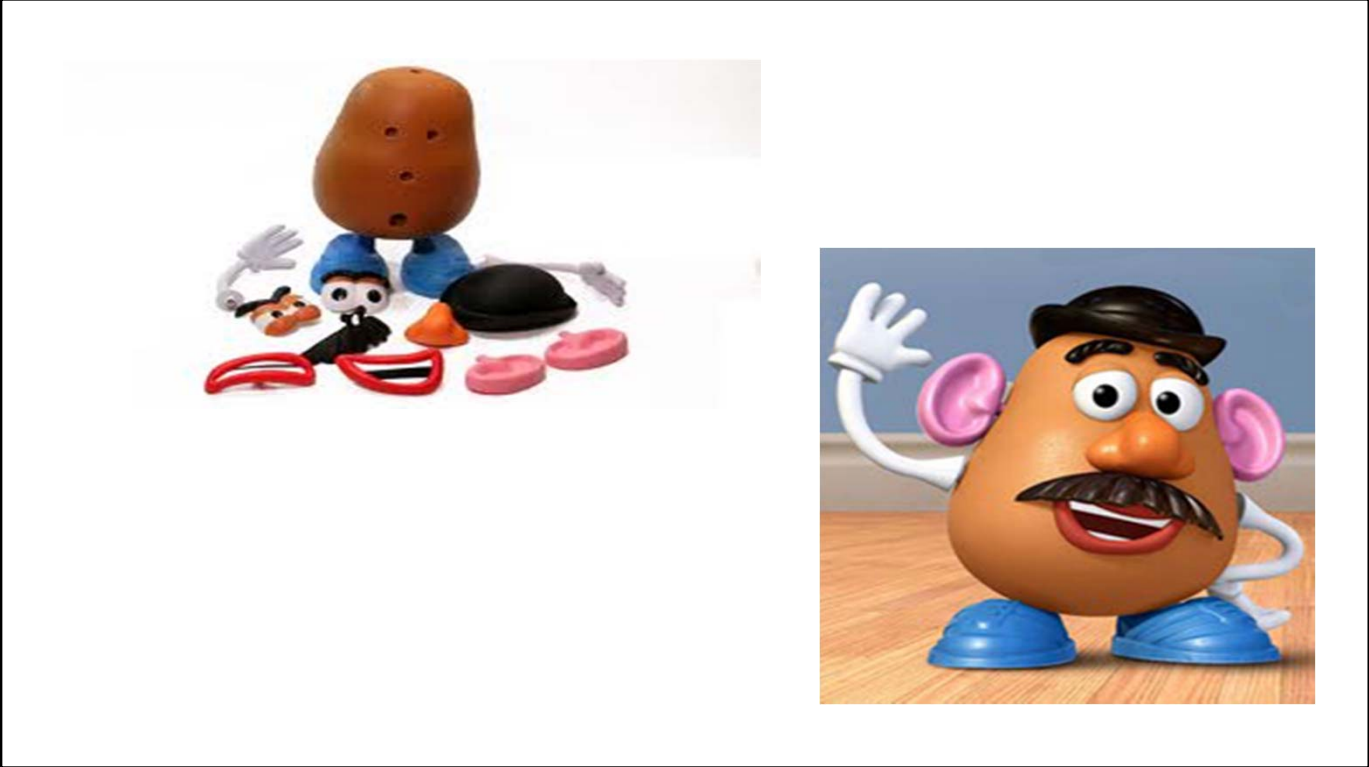


Decreased Cardiac Output





Ghaferi A, 2004  
Rychik, 2013.



**Indications**



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Preliminary Communications

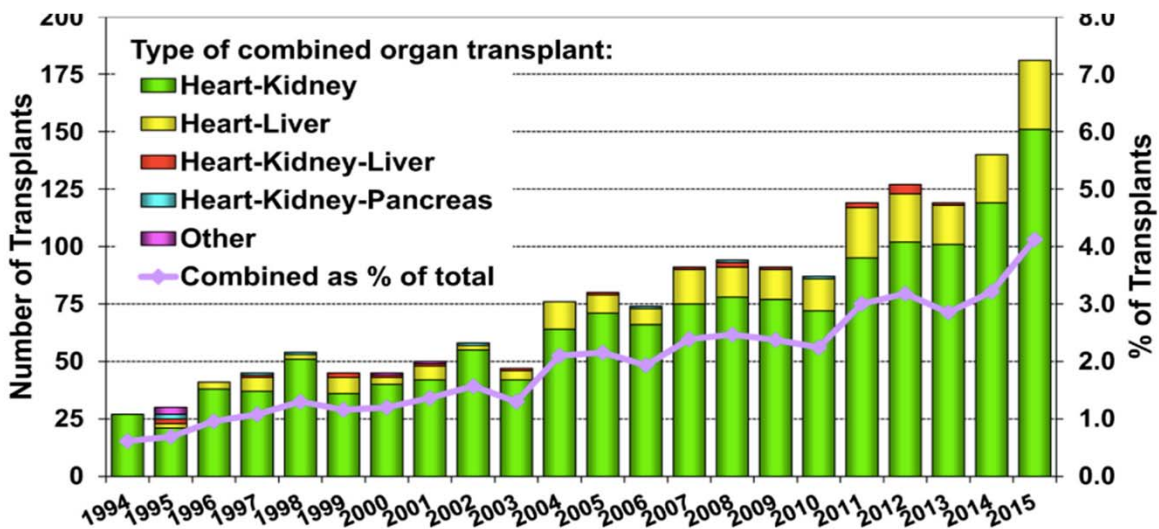
**HEART-LIVER TRANSPLANTATION  
IN A PATIENT WITH  
FAMILIAL HYPERCHOLESTEROLAEMIA**

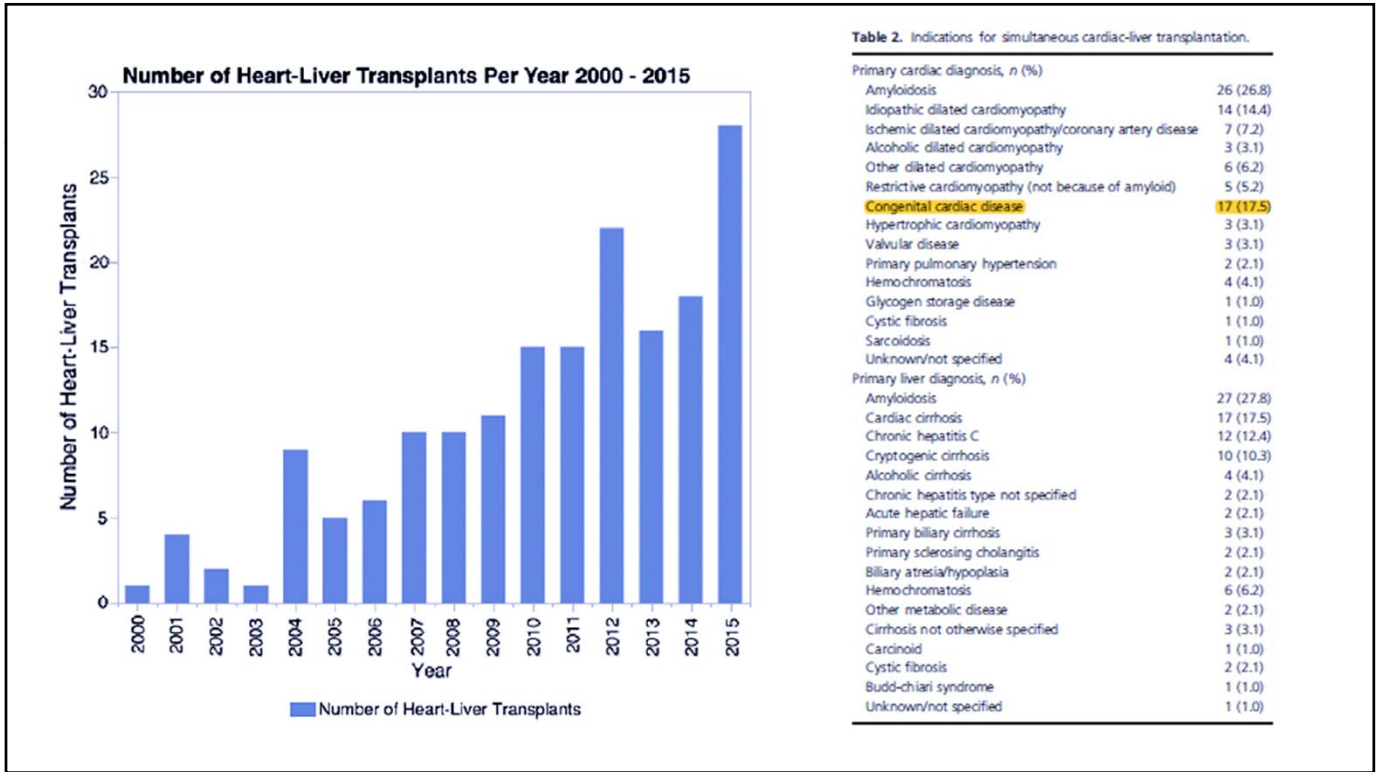
T. E. STARZL	D. W. BILHEIMER
H. T. BAHNSON	B. W. SHAW, JR
R. L. HARDESTY	B. P. GRIFFITH
S. IWATSUKI	B. J. ZITELLI
J. C. GARTNER, JR	J. J. MALATAK
A. H. URBACH	

*Departments of Surgery and Pediatrics, University of Pittsburgh Health Center, University of Pittsburgh, Pennsylvania; and Department of Internal Medicine, University of Texas Southwestern Medical School, Dallas, Texas, USA*



Multi-Organ Transplantation





**Table 2.** Indications for simultaneous cardiac-liver transplantation.

<b>Primary cardiac diagnosis, n (%)</b>	
Amyloidosis	26 (26.8)
Idiopathic dilated cardiomyopathy	14 (14.4)
Ischemic dilated cardiomyopathy/coronary artery disease	7 (7.2)
Alcoholic dilated cardiomyopathy	3 (3.1)
Other dilated cardiomyopathy	6 (6.2)
Restrictive cardiomyopathy (not because of amyloid)	5 (5.2)
<b>Congenital cardiac disease</b>	<b>17 (17.5)</b>
Hypertrophic cardiomyopathy	3 (3.1)
Valvular disease	3 (3.1)
Primary pulmonary hypertension	2 (2.1)
Hemochromatosis	4 (4.1)
Glycogen storage disease	1 (1.0)
Cystic fibrosis	1 (1.0)
Sarcoidosis	1 (1.0)
Unknown/not specified	4 (4.1)
<b>Primary liver diagnosis, n (%)</b>	
Amyloidosis	27 (27.8)
Cardiac cirrhosis	17 (17.5)
Chronic hepatitis C	12 (12.4)
Cryptogenic cirrhosis	10 (10.3)
Alcoholic cirrhosis	4 (4.1)
Chronic hepatitis type not specified	2 (2.1)
Acute hepatic failure	2 (2.1)
Primary biliary cirrhosis	3 (3.1)
Primary sclerosing cholangitis	2 (2.1)
Biliary atresia/hypoplasia	2 (2.1)
Hemochromatosis	6 (6.2)
Other metabolic disease	2 (2.1)
Cirrhosis not otherwise specified	3 (3.1)
Carcinoid	1 (1.0)
Cystic fibrosis	2 (2.1)
Budd-chiari syndrome	1 (1.0)
Unknown/not specified	1 (1.0)

419 Heart Transplants since 1977

7 Heart-Liver Transplants

Multi-Disciplinary Team

**BEST CHILDREN'S HOSPITALS**

**US NEWS**

RANKED IN 10 SPECIALTIES 2018-19

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## Our First 7 Cases



7 Patients (5 Girls, 2 Boys)

Ages 12-19

6 Single Ventricles with FALD, 2 with PLE

One TOF (2 ventricle) with primary liver disease



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## Heart-Liver Transplant Planning



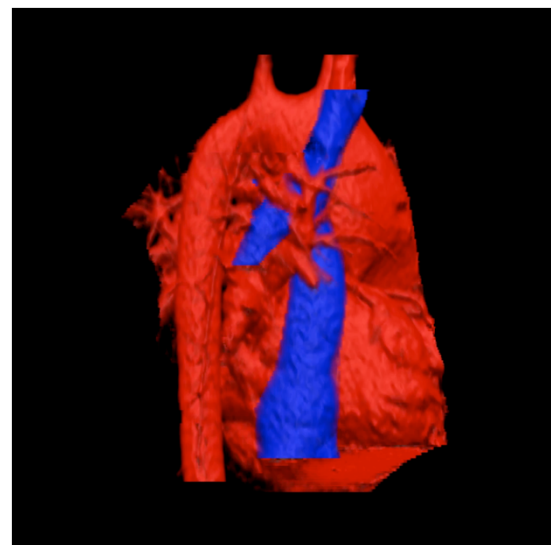
Close collaboration between heart and  
liver teams

Standardized multidisciplinary work-up.

Ultrasound, CT, MRI

Cardiac Catheterization/Hepatic Vein Wedge  
Pressure Gradient

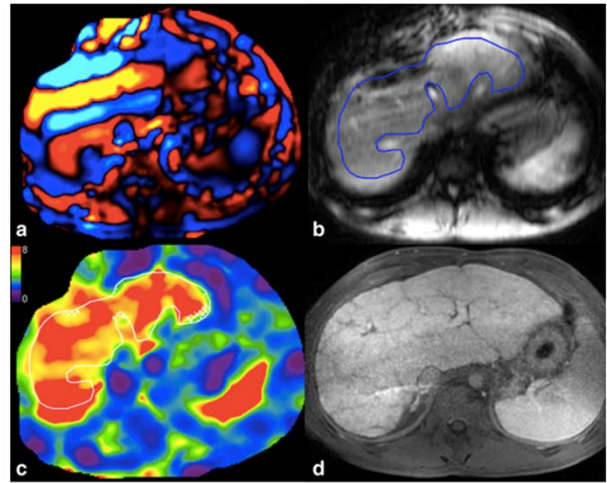
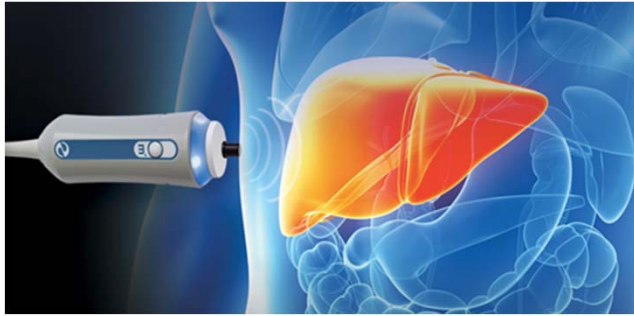
Consider liver biopsy (during cath)



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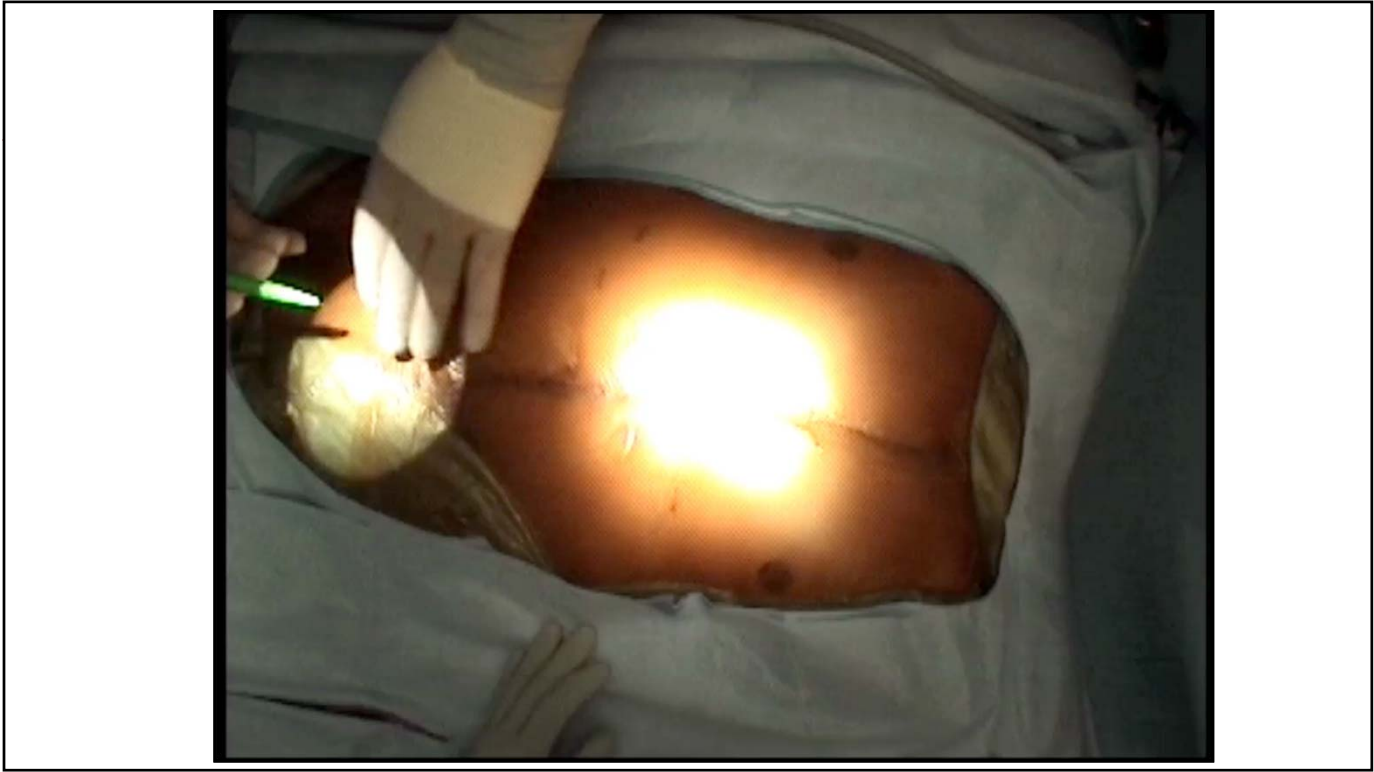
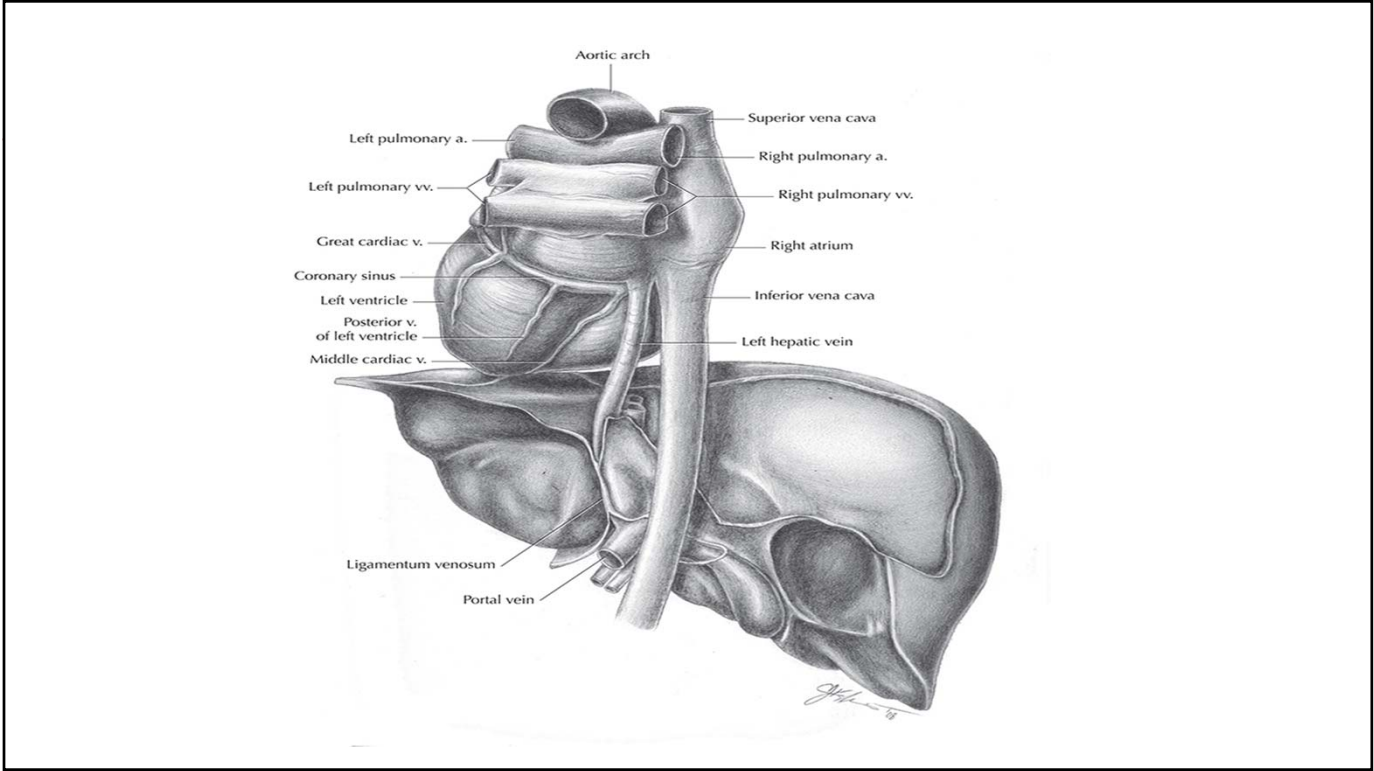
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# Elastography



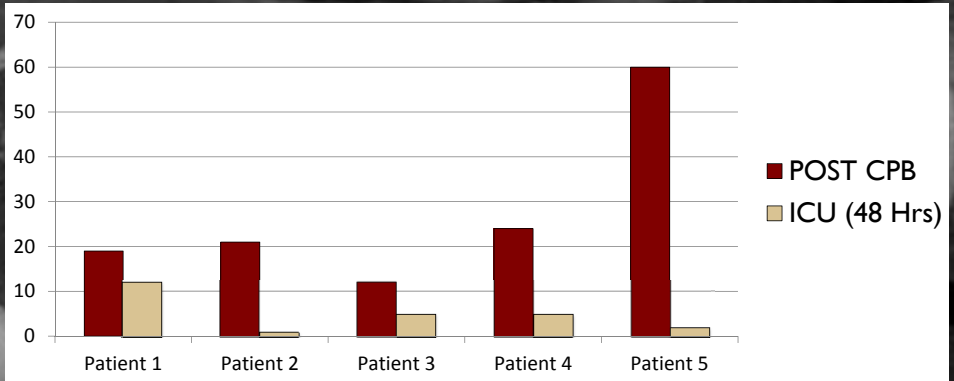
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Waitlist time: **131 Days (50-622)**  
CPB: **3.58 Hours (2.97- 4.15)**  
Donor Ischemic Time: **4.13 Hours (3.78-6.6)**  
Total OR Time: **(13-18.65)**



## Transfusions in the OR



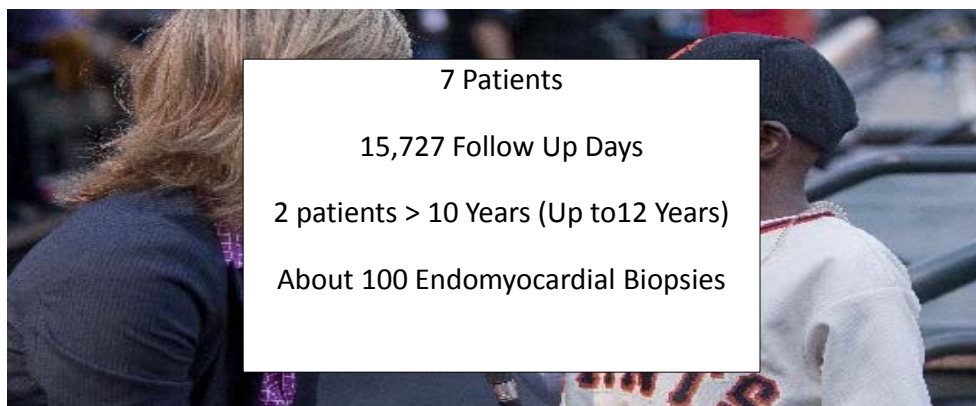
	MEAN	MEDIAN	RANGE
pRBC (units)	22.2 ( $\pm$ 10.3)	19	14-40
FFP (units)	17.6( $\pm$ 15.3)	10	5-42
Platelets (units)	4 ( $\pm$ 3.5)	3	1-10
Cryoprecipitate (10 pack unit)	2.2 ( $\pm$ 1.3)	2	1-4



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## How do they do?



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## Where are they now?



All are alive and well

Normal Cardiac and Liver Function

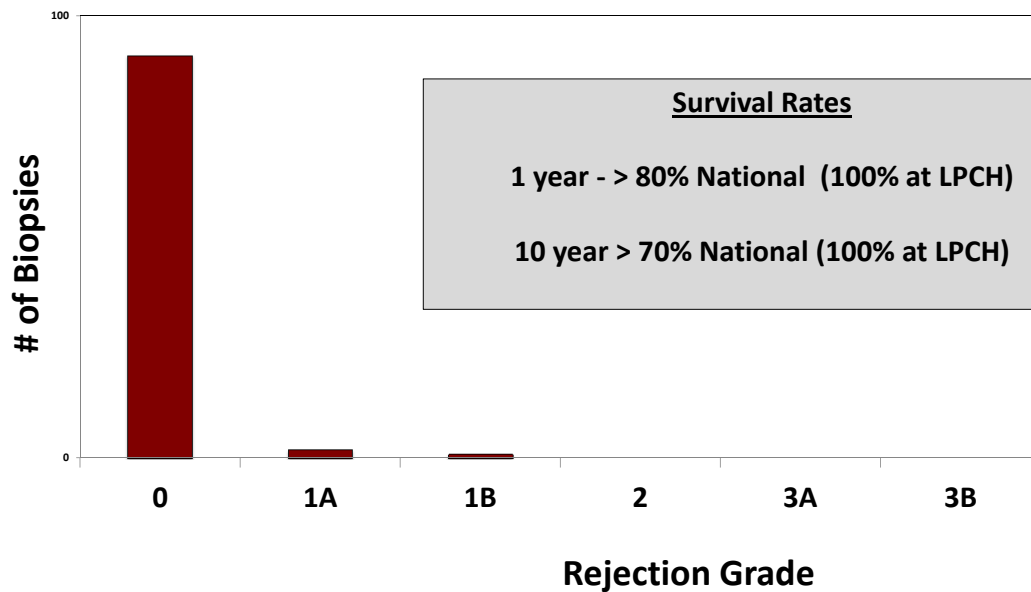
3 Transitioned to adult programs

*But most impressively....*



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## Rejection Rates





# Why do they do so well?



More immunosuppression?  
More surveillance?  
More biopsies?



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## Immunologic Tolerance

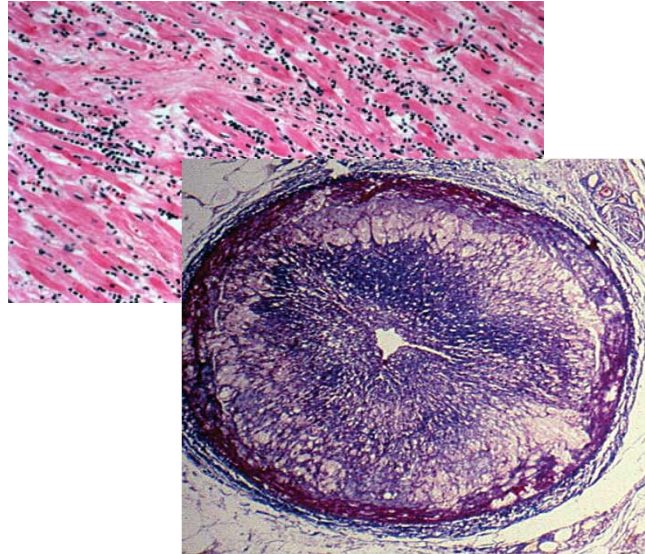


Liver protects companion organ in many combinations.

Reduced rejection of both organs.

Reduced incidence of coronary vascular disease

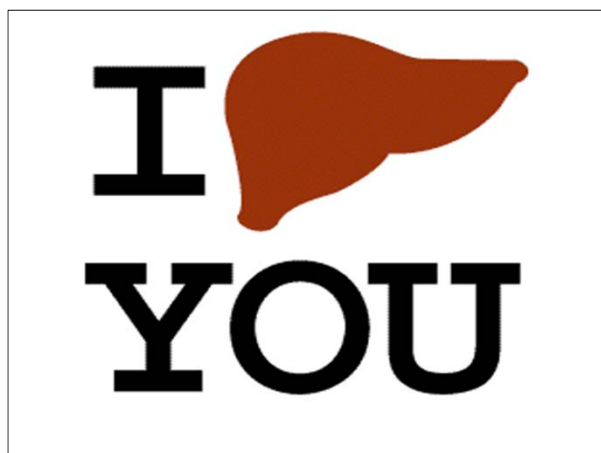
Need for less immunosuppression.



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## Immunologic Tolerance



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## UNOS Final Rule



The organ allocation system in the US is to prioritize patients with the greatest medical urgency while at the same time maximizing the overall societal benefit from a limited pool of organs.

Also takes into account the change in quality of life, duration of benefit, and *amount of resources required* for successful treatment.

## Our obligation as stewards...



To prioritize patients with the greatest medical urgency

... jumping the line for the 2<sup>nd</sup> organ?

...at the same time maximizing the overall societal benefit from a limited pool of organs.

... saving one life instead of 2?

Also takes into account the change in quality of life, duration of benefit, and *amount of resources required* for successful treatment.



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## Commitment to Patient vs. Society



Individual  
Patient Advocacy



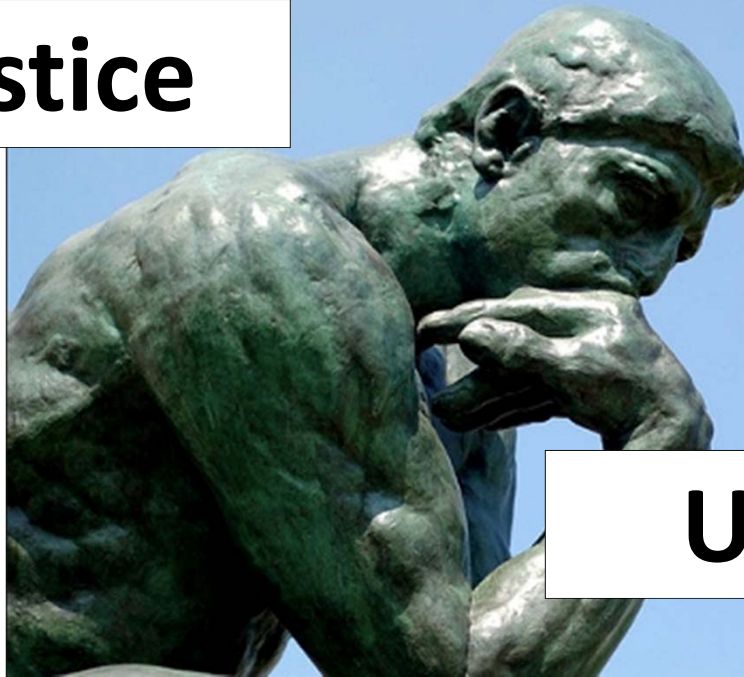
Greater Good



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# Justice



# Utility

## Conclusions



Heart-Liver Transplant is increasing in frequency in adults and children

Adolescents and adults with congenital heart disease make up a significant proportion of heart-liver transplant candidates

Indications are unclear

Heavy surgical burden but patients survive

Long-term prognosis is excellent



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