

# Hypoplastic left heart syndrome Hypoplasie du cœur gauche



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Cardiopathies congénitales humaines	Fréquence	Incidence
Communication interventriculaire (CIV)	30%	1500
Communication interauriculaire (CIA)	8%	400
Sténose pulmonaire (SP)	7%	350
Persistance du canal artériel (PCA)	7%	350
Coarctation de l'aorte (CoA)	6%	300
Tétralogie de Fallot (T4F)	6%	300
Transposition des gros vaisseaux (TGV)	5%	250
Sténose aortique (SA)	5%	250
Canal atrioventriculaire (CAV)	4%	200
<b>Syndrome d'hypoplasie du cœur gauche (SHCG)</b>	<b>3.5%</b>	<b>180</b>
Atrésie pulmonaire à septum intact (APSI)	2%	100
Atrésie pulmonaire à septum ouvert (APSO)	2%	100
Atrésie tricuspide (AT)	2%	100
Tronc artériel commun (TAC)	2%	100
Retour veineux pulmonaire anormal (RVPA)	2%	100
Malpositions vasculaires (MV)	1%	50
Interruption de l'arc aortique (IAA)	1%	50
Ventricule unique (VU)	1%	50
Anomalie d'Ebstein	1%	50
Discordances AV et VA	1%	50
Autres	6%	300

# HLHS: définition

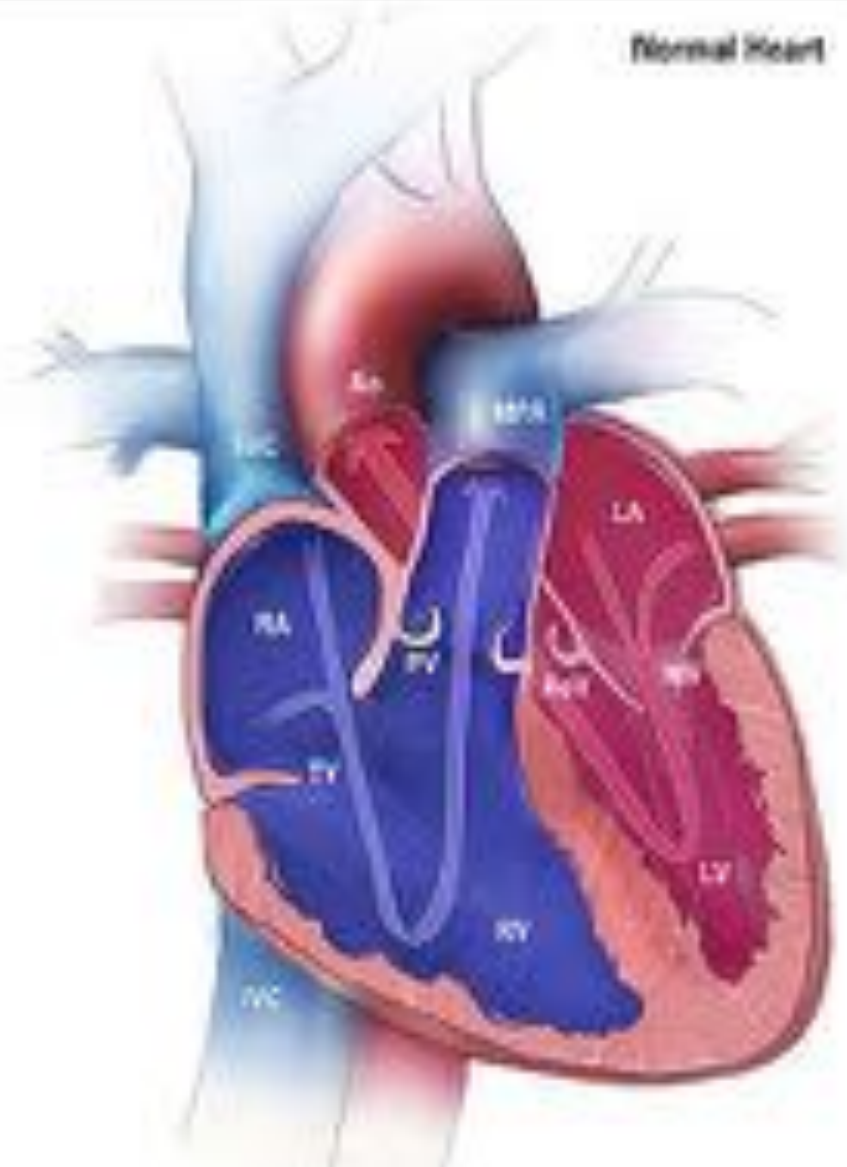
Dépendance de la circulation systémique du canal artériel à cause d'une atrésie ou hypoplasie mitro-aortique avec hypoplasie du ventricule gauche

Situs solitus

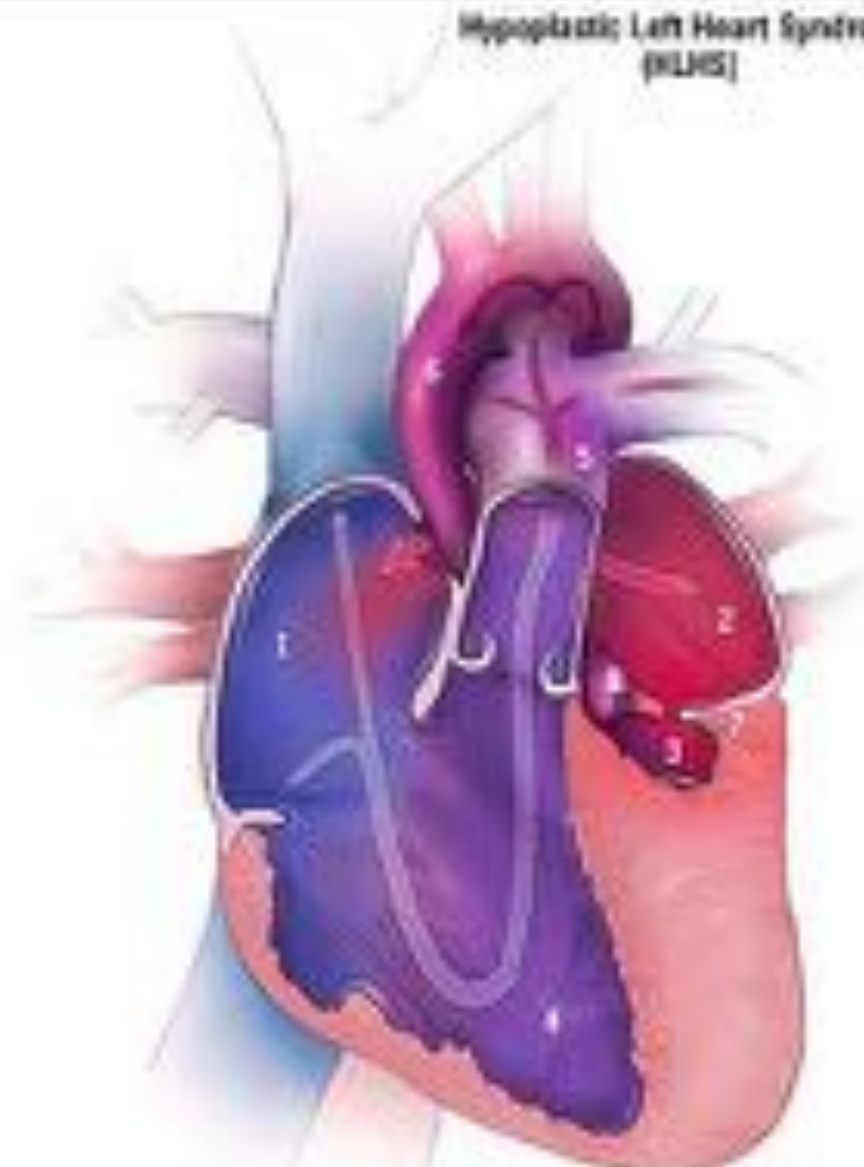
D-loop ventriculaire

Concordance AV et concordance VA

Normal Heart



Hypoplastic Left Heart Syndrome (HLHS)



**Chambers of the Heart**  
 RA, Right Atrium  
 LA, Left Atrium  
 RV, Right Ventricle  
 LV, Left Ventricle

**Vessels**  
 PA, Main Pulmonary Artery  
 Ao, Aorta  
 SVC, Superior Vena Cava  
 IVC, Inferior Vena Cava

**Valves**  
 TV, Tricuspid Valve  
 MV, Mitral Valve  
 AV, Aortic Valve  
 PV, Pulmonary Valve

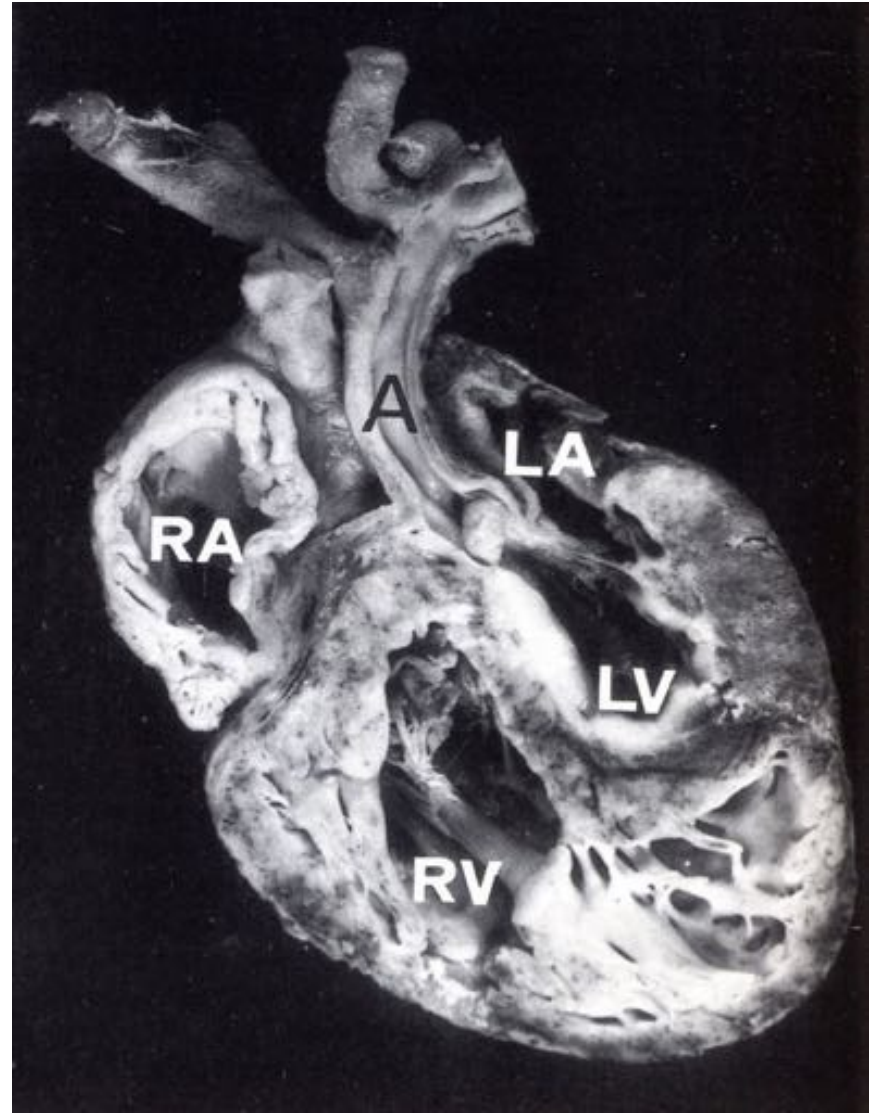
1. Right Atrium  
 2. Left Atrium  
 3. Left Ventricle  
 4. Right Ventricle  
 5. Pulmonary Artery

6. Aorta  
 7. Mitral Valve  
 8. Aortic Valve

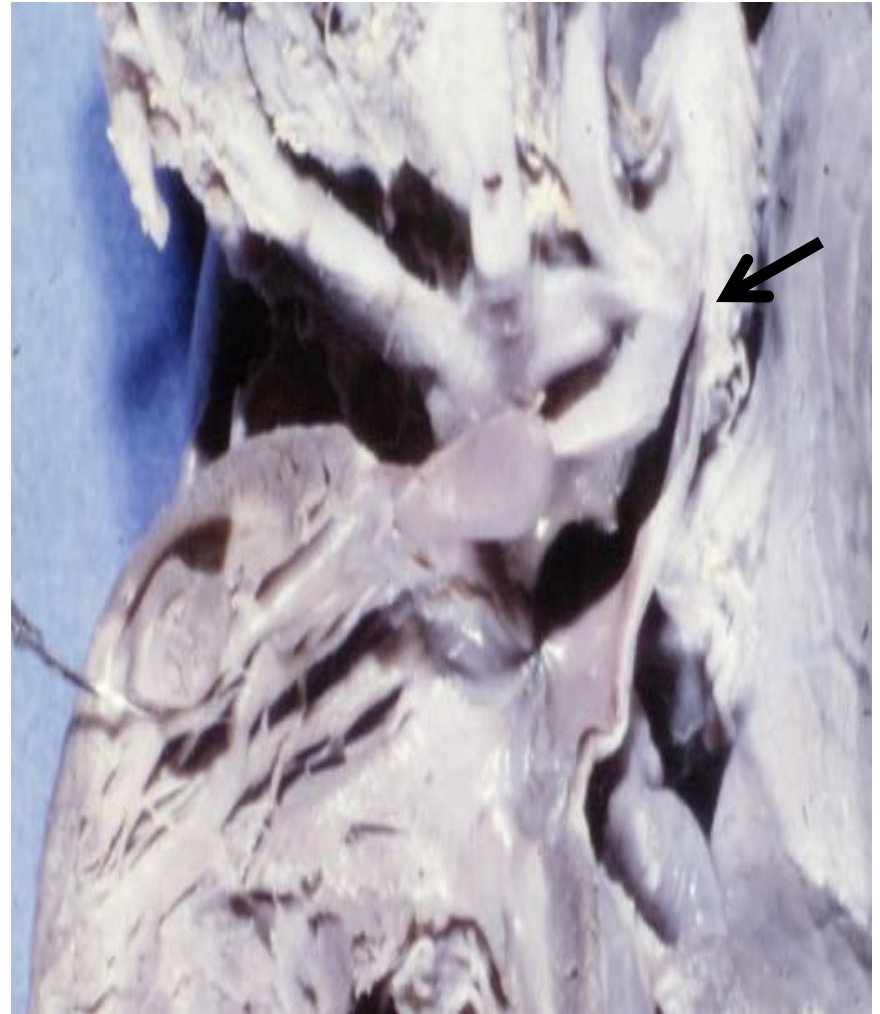
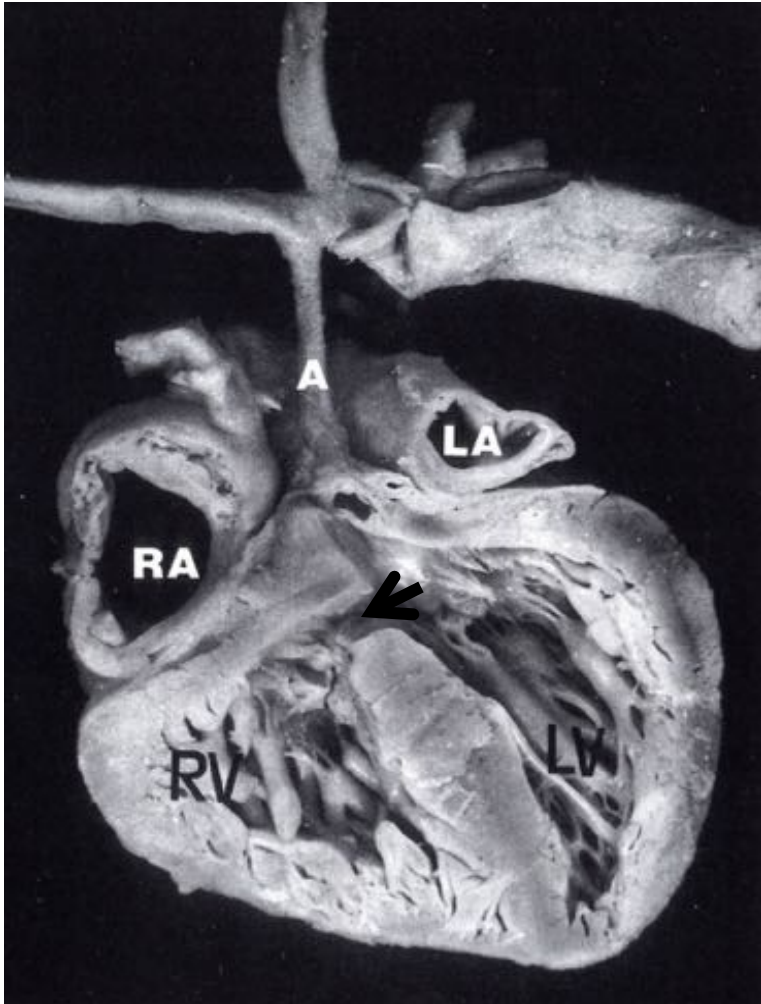
# HLHS: anatomie



# HLHS: anatomie



# HLHS: anatomie



# HLHS: anatomie particulière

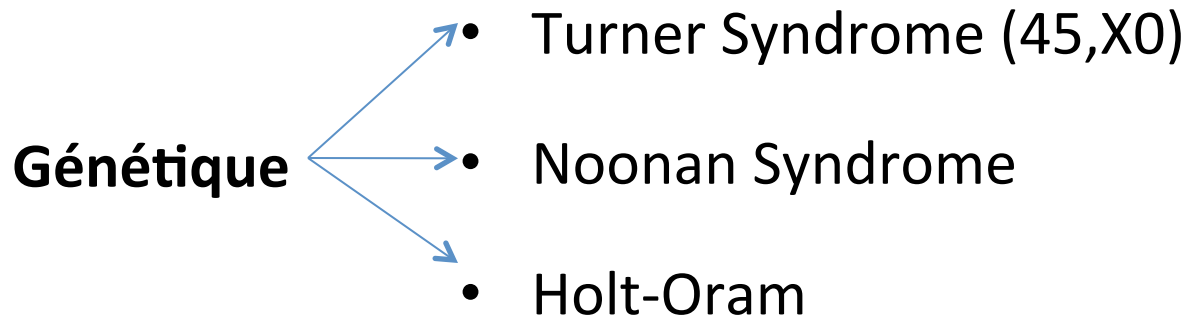
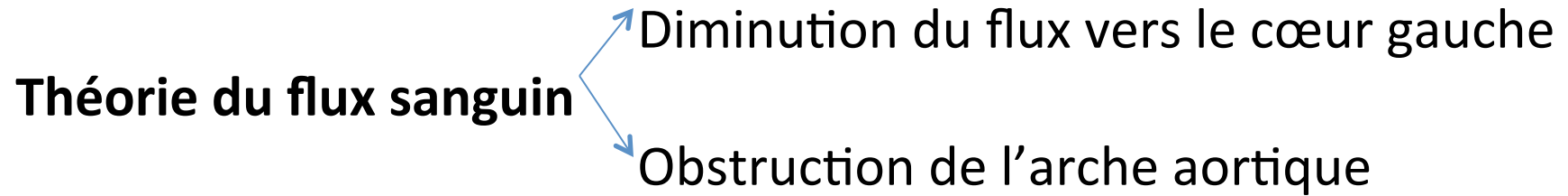
- CIV associée
- 5 % anomalies RVS – le plus souvent VCSG
- 5% anomalies RVP – RVPAT , veine lévocardinale au TVI
- Absence de CIA ou CIA restrictive
- Fistules coronaro-camérales – rares
- ALCAPA (case reports)



# HLHS: epidemiologie

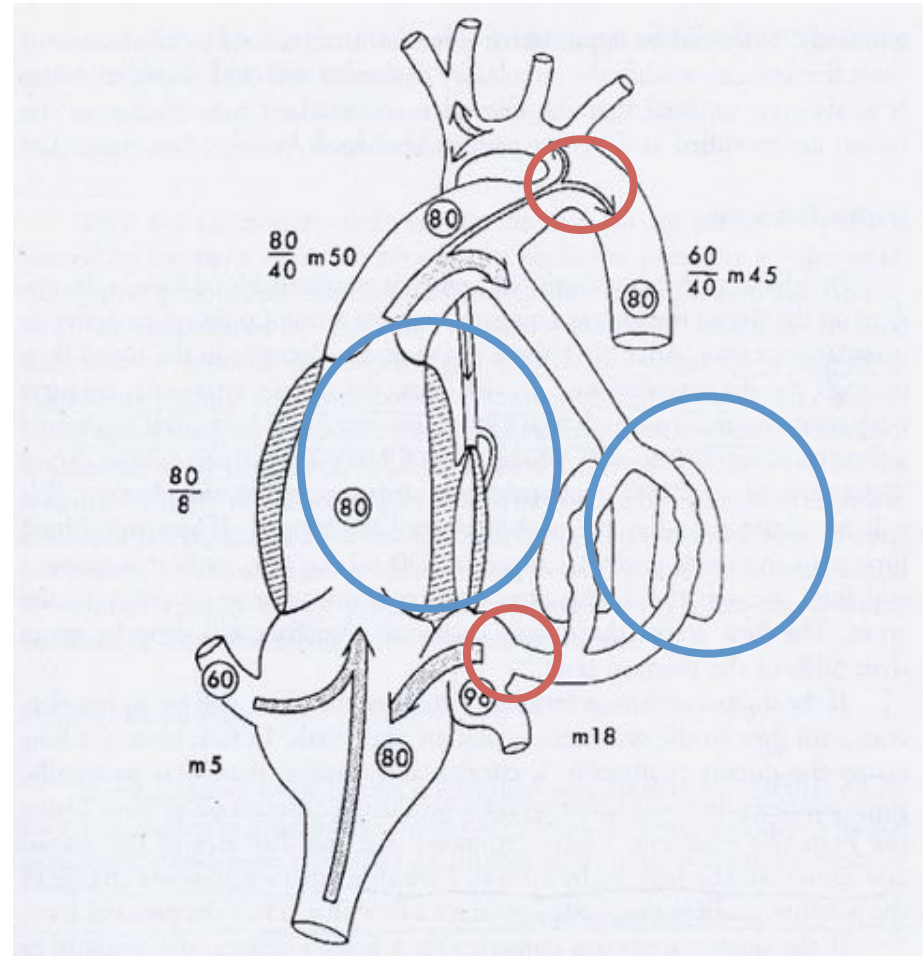
- **Prévalence** 0,2-0,3/1000 naissances  
3,5% de CC - 55% masculins
- **Histoire naturelle** 95% mortalité pendant le 1er mois  
80% mortalité pendant la 1ère semaine
- **Récurrence familiale** 0,5-2% dans la fratrie
- **Extracardiaque** Chromosomal 10%  
Malformation majeure 10%  
Autre 10%

# HLHS: morphogenese-génétique



# HLHS: hémodynamique

- Shunt G-D obligatoire
- Hypoxémie aortique
- $PO_2$  élevée dans l'AP
- Flux critique : CA
- Role des RVP
- Flux critique: CIA
- Fonction VD systémique



# HLHS: diagnostic anténatal

## Etude EPICARD

### ii) Hypoplastic Left Heart Syndrome

	1983 - 1988			1989 - 1994			1995 - 2000			
	N	%	95 % CI*	N	%	95 % CI*	N	%	95 % CI*	p <sup>‡</sup>
Prenatal Diagnosis	22	31.8	13.9 - 54.9	29	82.8	64.2 - 94.2	27	88.9	70.8 - 97.6	< 0.001
Pregnancy Termination	22	13.6	2.9 - 34.9	29	72.4	52.8 - 87.3	27	63.0	42.4 - 80.6	< 0.001
First Week Mortality	18	83.3	58.6 - 96.4	8	75.0	34.9 - 96.8	10	50.0	18.7 - 81.3	0.12
Perinatal Mortality	19	84.2	60.4 - 96.6	8	75.0	34.9 - 96.8	10	50.0	18.7 - 81.3	0.10



Heart. 2012 Aug 11. Prevalence, timing of diagnosis and mortality of newborns with congenital heart defects: a population-based study. Khoshnood B et al

# Impact of Prenatal Diagnosis in Survivors of Initial Palliation of Single Ventricle Heart Disease

Analysis of the National Pediatric Cardiology Quality Improvement Collaborative Database

Brown et al. 2015 Ped Cardiol

- National Pediatric Cardiology Quality Improvement Collaborative initiée en 2008 aux USA
- Réseau de 50 centres participant; **n=591** (06/2008-06/2012)

**Table 2** Comparison of patient characteristics by prenatal cardiac diagnosis (PD, n = 591)

Baseline patient information	PD, n = 445 median [IQR], or number (%)	No PD, n = 146 median [IQR], or number (%)	p value
Birth weight (kg) [n = 443, 141]	3.1 [2.9,3.5]	3.2 [2.9, 3.5]	0.72
Gestational age (weeks) [n = 439, 146]	39 [38,39]	39 [38,40]	<0.001
APGAR score—1 min [n = 436, 125]	8 [8,8]	8 [8,9]	0.15
APGAR score—5 min [n = 436, 125]	9 [8,9]	9 [9,9]	<0.001
Age at presentation (days)	0 [0,0]	1 [0,4]	<0.001
Lowest pH prior to surgery	7.35 [7.35,7.35]	7.35 [7.35,7.35]	0.35
Primary cardiac diagnosis (n = 444,145)			0.53
Age at admission to ICU (days)	0 [0,0]	1 [0,4]	<0.001
Age at initial intubation (days) [n = 442,146]	3 [0,5]	3 [1,8]	0.02
Any pre-operative risk factors	218 (48 %)	99 (68 %)	<0.001
Mechanical ventilatory support	128 (29 %)	75 (51 %)	<0.001
Acidosis	61 (14 %)	49 (34 %)	<0.001
Renal insufficiency	22 (5 %)	17 (12 %)	0.007
Arrhythmia	17 (4 %)	3 (2 %)	0.43
Shock, resolved, or persistent	1 (<1 %)	3 (2 %)	0.05
AV valve regurgitation <sup>a</sup>	7 (2 %)	10 (7 %)	0.002
Ventricular dysfunction <sup>a</sup>	8 (2 %)	7 (5 %)	0.06

Pas d'impact sur la mortalité interstage ni résultat DCCP

# Sténose aortique: intervention foétale

- Bénéfice théorique pour permettre une réparation biventriculaire ?
- Score **eHLHS** «evolving HLHS» pour identifier les foetus avec St. aortique avec un potentiel d'évoluer vers l'HypoVG (Maekikallio et al. 2006 Circ)
- «**threshold score**» pour foetus avec sténose aortique et score eHLHS > 3: si élevé peu/pas de chance d'obtenir une circulation biventriculaire

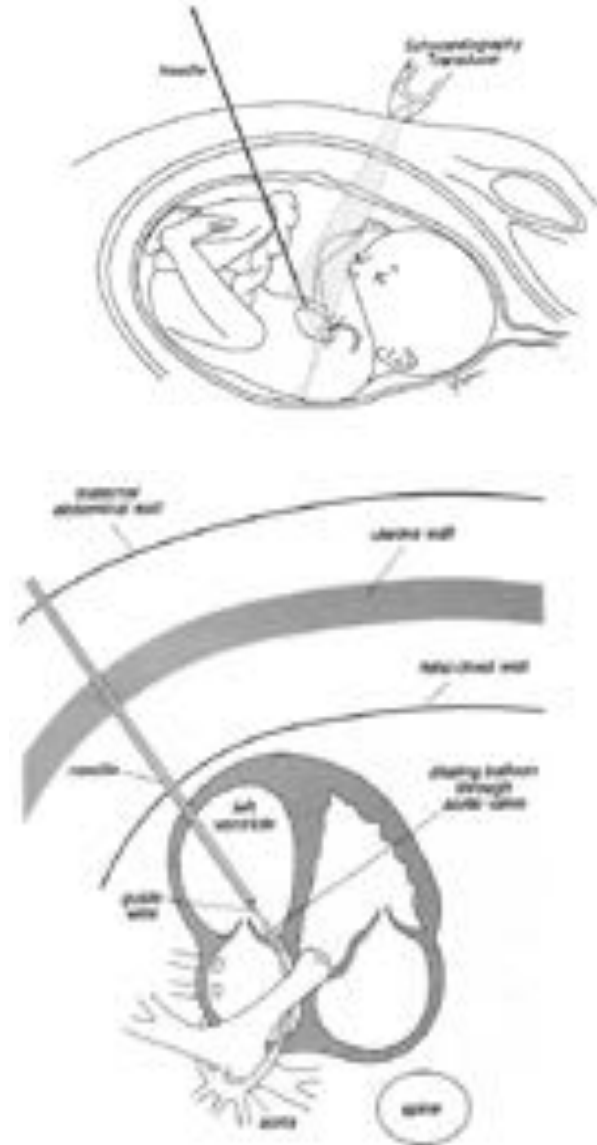
(McElhinney et al. 2009, Circ)

# Déroulement de l'intervention

## Procédure

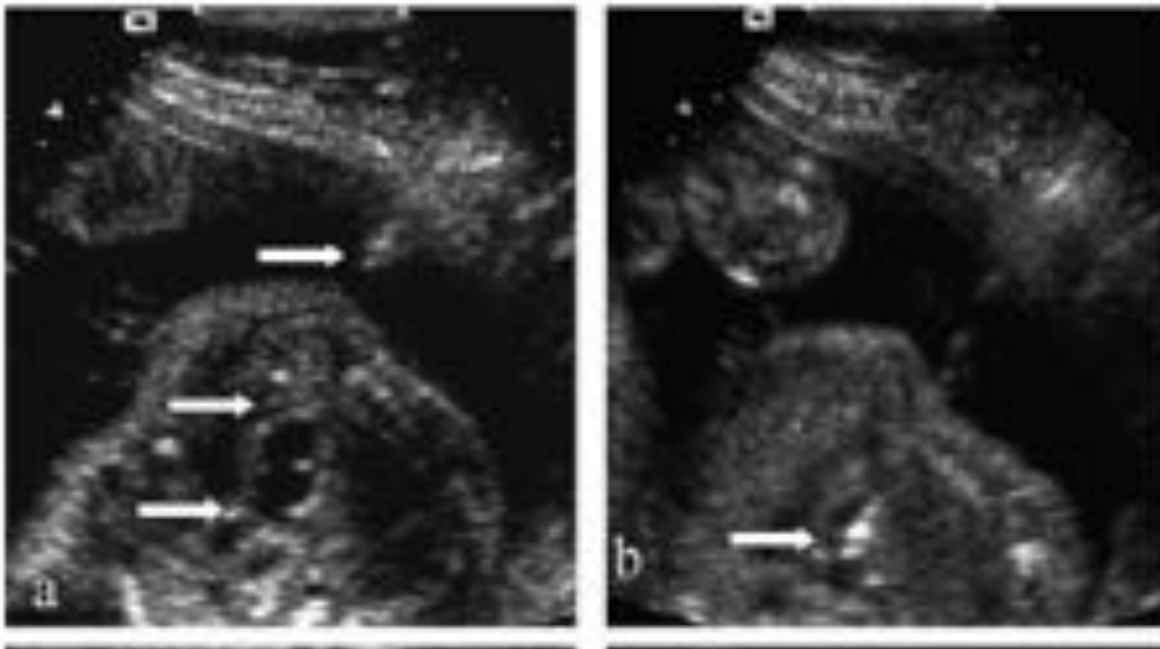
- Anesthésie maternelle: locale
- 1<sup>ère</sup> étape: anesthésie générale du du fœtus
- Ponction écho guidée
- Aiguille 18 Gauge
- Ratio diamètre du ballon/anneau aortique 1/1
- Inflation à haute atmosphère pour avoir un ratio proche de 1,2
- Déflation du ballonnet
- Retrait de l'aiguille
- Contrôle écho: péricarde, fonction VG, fuite aortique

Expérience NEM  
Courtesy of Y Boudjemline



# Problèmes techniques

- Après ponction dégradation de la fonction VG: procédure rapide+++
- Procédure traumatique:
  - Hémopéricarde constant



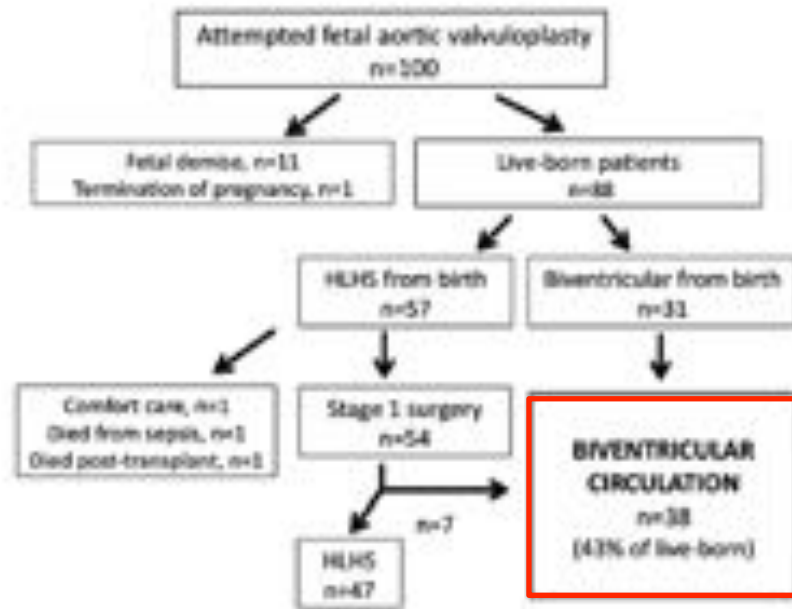
Expérience NEM  
Courtesy of Y Boudjemline



# Fetal Aortic Valvuloplasty for Evolving Hypoplastic Left Heart Syndrome

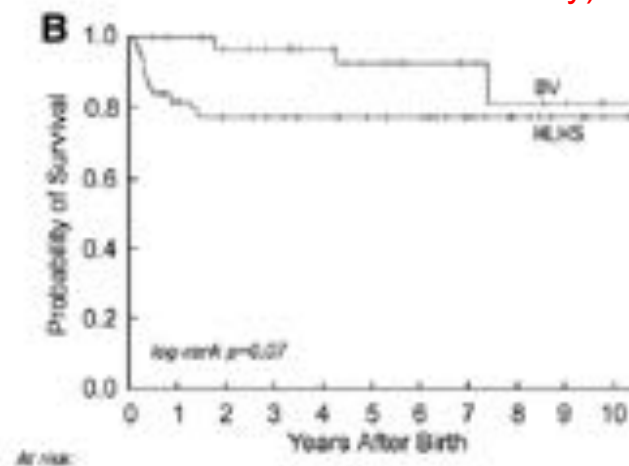
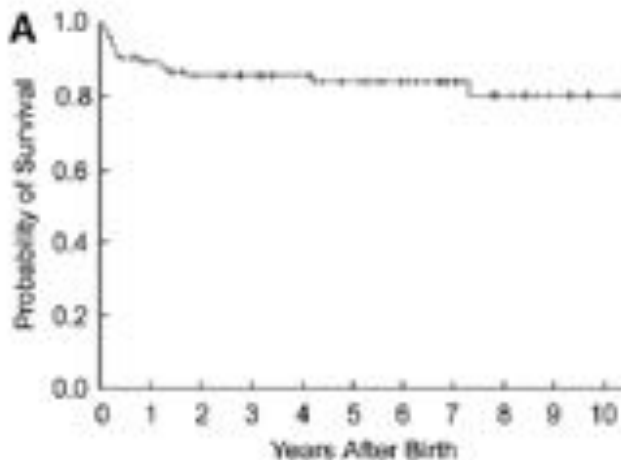
## Postnatal Outcomes of the First 100 Patients

03/2000-01/2013



15 pts (39%):  
replacement aortique  
(Ross, mécanique)

8 pts (21%):  
replacement mitral  
Bioprothese, mécanique,  
melody)



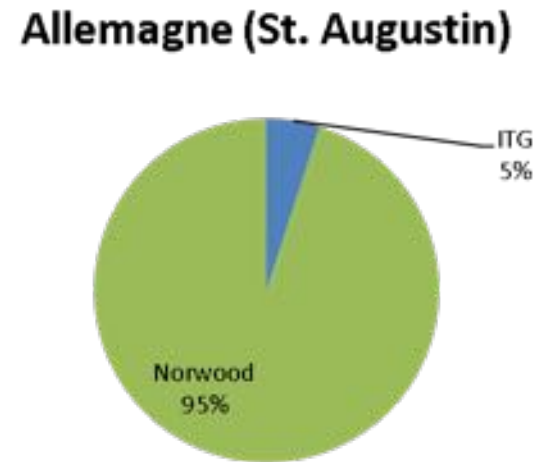
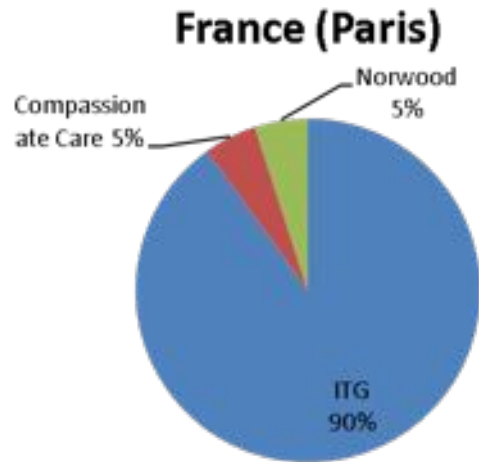
# HLHS: Conseil parental

- Interruption thérapeutique de grossesse
- Accompagnement à la naissance = compassionate care
- Traitement palliatif chirurgical en 3 étapes avec l'objectif d'une circulation de Fontan (Norwood/ Sano/ Hybrid Stage 1 puis DCPP/ DCPT)

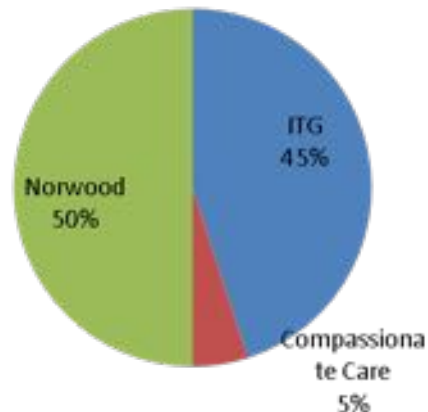
# HLHS: Conseil parental

- Morbidité-mortalité d'une intervention chirurgicale complexe en période néonatale
- Morbidité-mortalité en attendant Stage 2
- Qualité de vie à long terme, devenir neurologique...

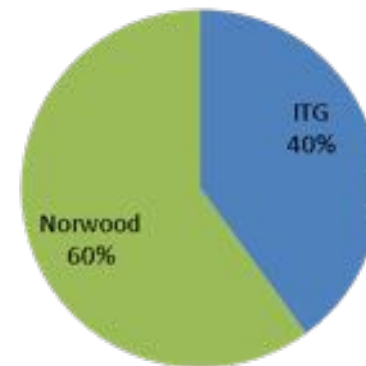
# HLHS: Décision parentale: DAN



**Angleterre (Londres)**

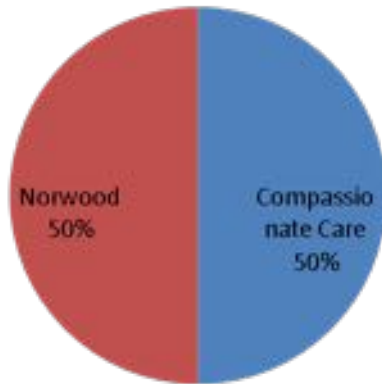


**Belgique (Leuven)**

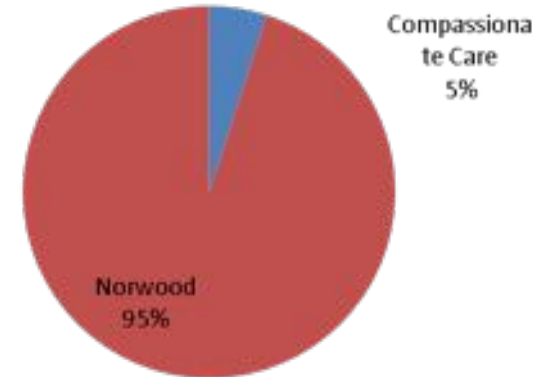


# HLHS: Décision parentale: DPN

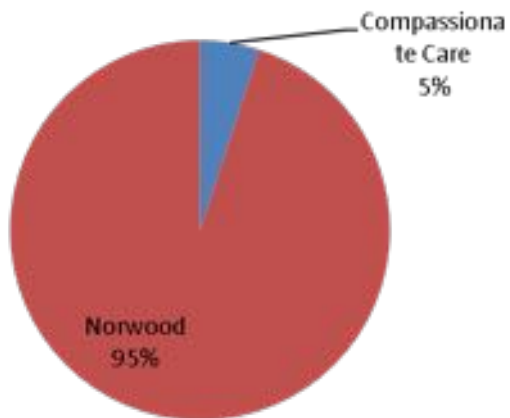
France (Paris)



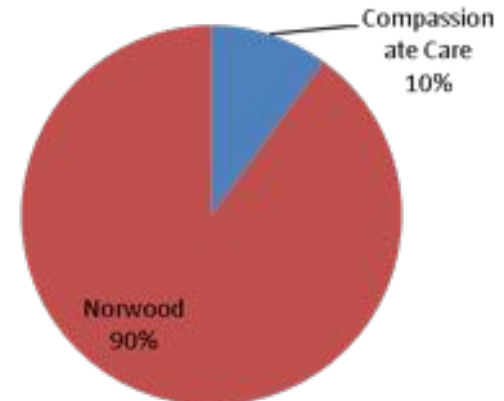
Allemagne (St. Augustin)



Angleterre (Londres)



Belgique (Leuven)



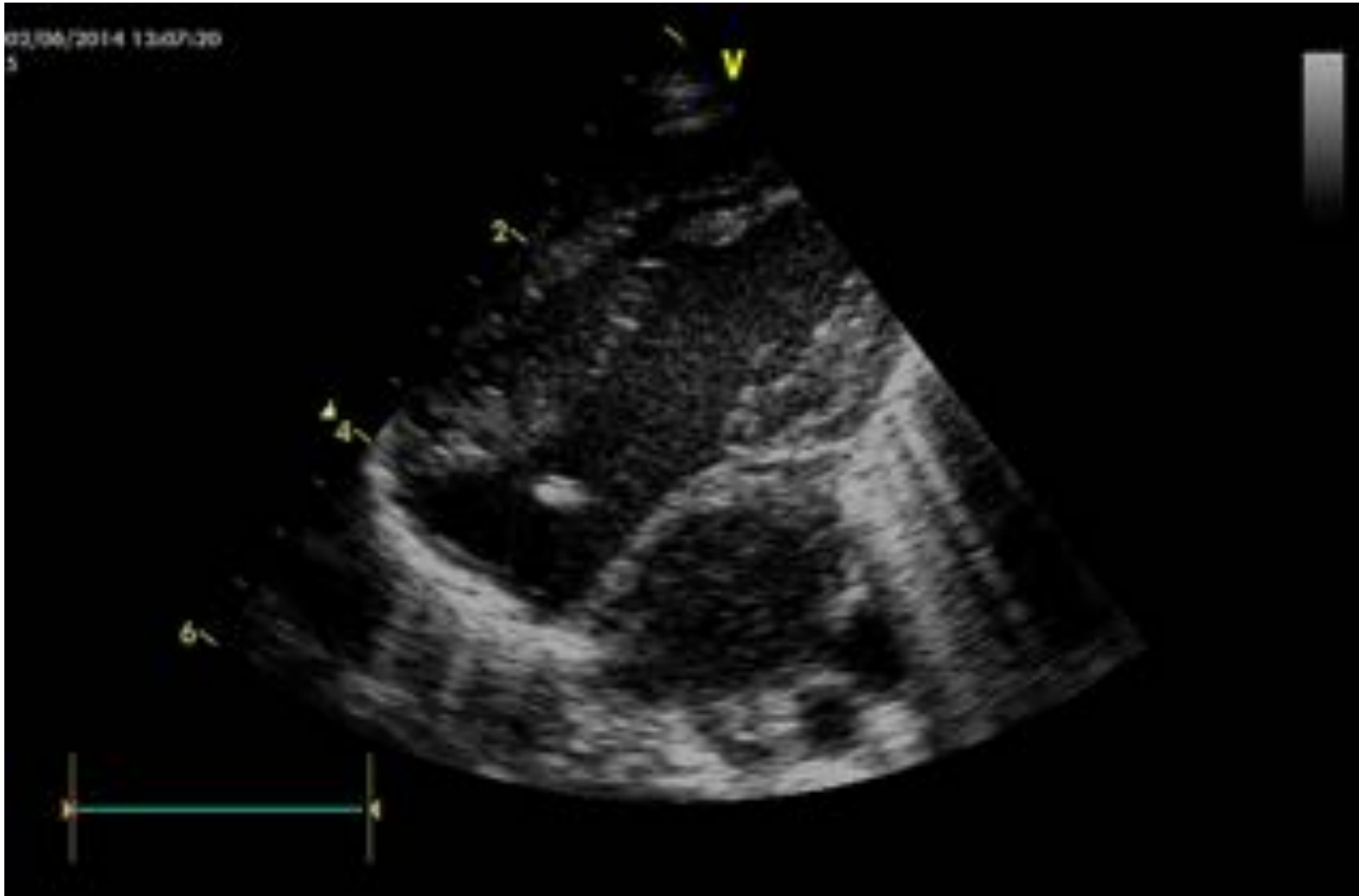
# HLHS: diagnostic clinique

## **Tableau d'insuffisance cardiaque:**

- Cyanose modérée et diminution des pouls sans souffle cardiaque
- Etat de choc multi viscérale à la fermeture du CA
- OAP avec dyspnée et cyanose marquée si CIA restrictive

25% des enfants sont symptomatiques dans les 24 hrs

# HLHS: diagnostic ETT

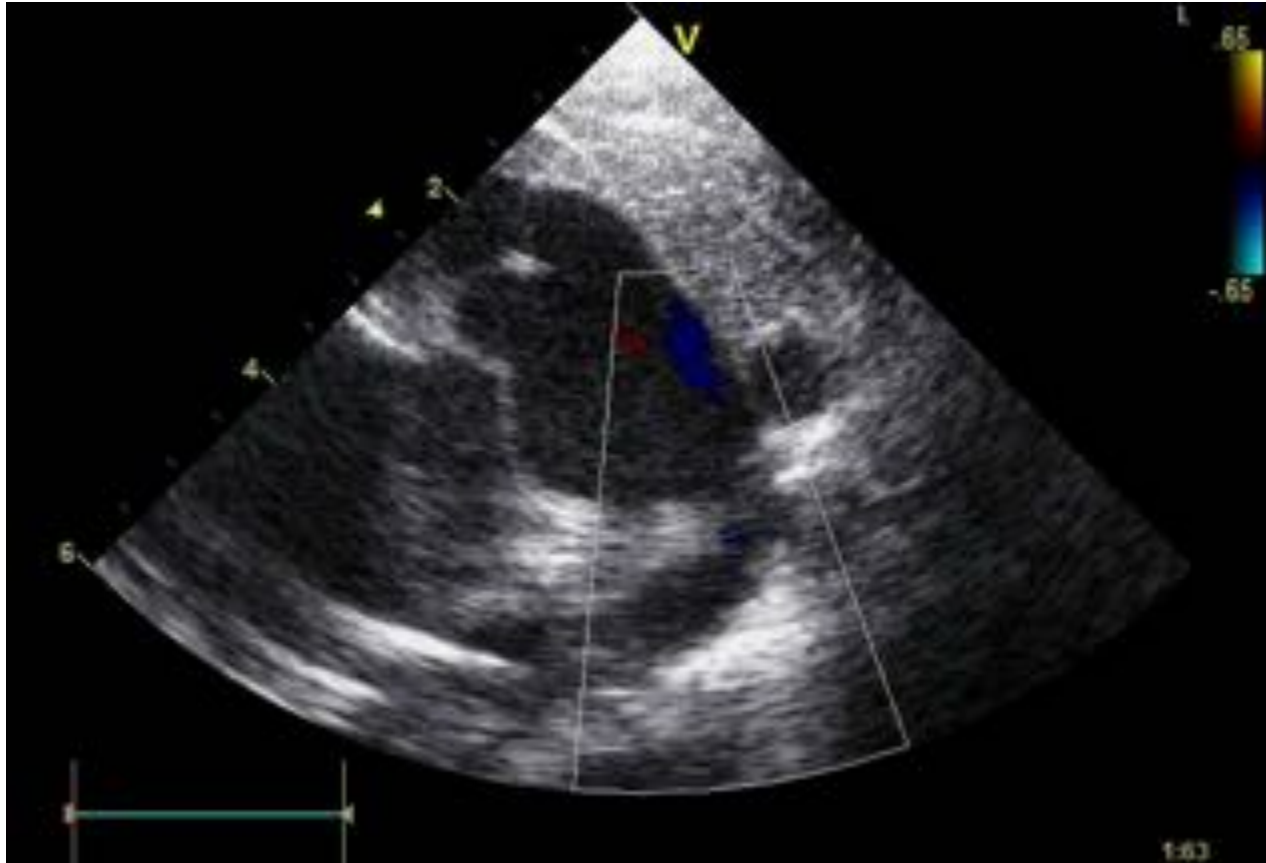


# HLHS: diagnostic ETT





# HLHS: diagnostic ETT



# Evaluation échographique pré-op

- Forme anatomique HLHS
- Fonction du VU
- Dysplasie/fuite tricuspide
- RVP normal ou anormal?
- Taille de la CIA: à agrandir (RSK, atrioseptectomie)
- Fistules coronaires ?
- Taille de l'aorte ascendante, coarctation

# HLHS: High risk – high mortality

- **Facteurs généraux:** prématurité, hypotrophie, anomalie extracardiaque, anomalie chromosomique
- **Facteurs cardiaques:** dysfonction VD, fuite tricuspide modérée ou importante, retour veineux anormal total, CIA restrictive, petite taille de l'aorte ascendante
- 37% de survie en cas de FdR et DAN HypoVG vs. 79% sans FdR +DAN \*

\*Rychik et al. 2010 Ultrasound Obstet Gynecol

# Evaluation échographique post-op

- Fonction VU
- Fuite tricuspide
- Perméabilité du BT-Shunt ou Sano ou stent
- Vitesse des cerclages si procédure hybride
- CIA non véloce si stent ou RSK
- Absence de CoA

# PEC médical post-opératoire

- Nutrition optimal (+/- SNG)
- +/- Diurétiques
- IEC si dysfonction VU ou fuite
- Aspirine pour Blalock/Stent/Sano
- Synagis en hiver
- Follow-up rapproché en consultation pour dépister des complications, la majoration de la cyanose, stagnation pondérale

# Interstage mortality

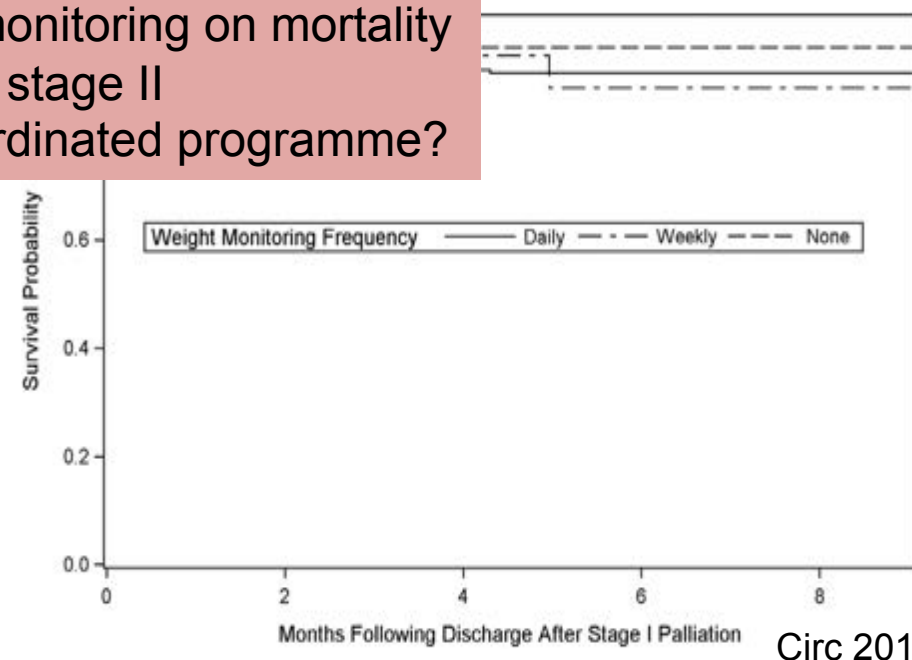
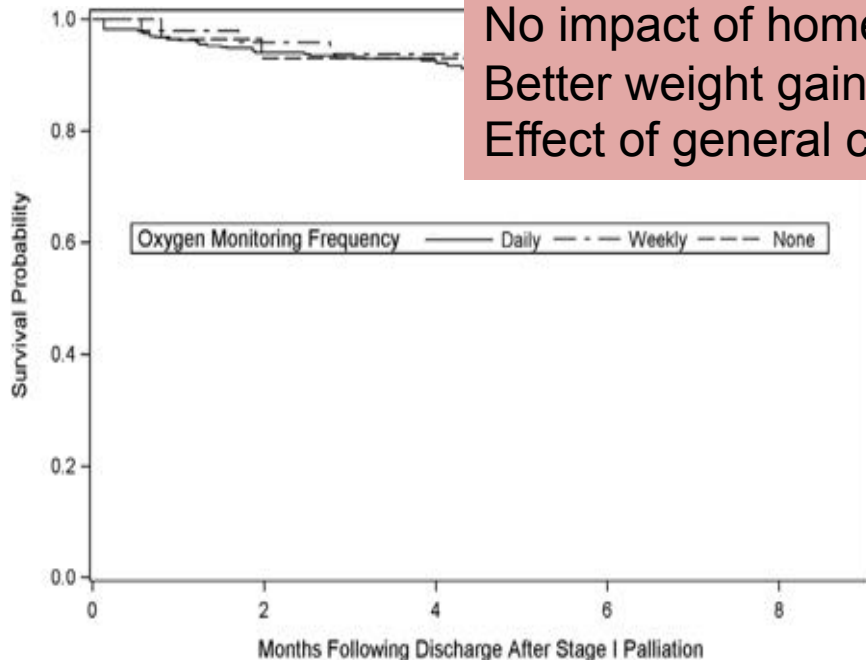
N= 509

Oster et al Outcomes of Interstage Home Monitoring

**Table 2. Association of Interstage Home Oxygen Saturation Monitoring Frequency With Outcomes for Infants With Single-Ventricle Heart Disease**

	Daily (Referent; n=397), n (%)	Weekly (n=61) n (%)	Adjusted OR* (95% CI)	PValue	None (n=36)	Adjusted OR* (95% CI)	PValue
Interstage mortality	34 (9)	4 (7)	1.10 (0.35–3.50)	0.87	2 (6)	0.85 (0.19–3.79)	0.83
Interstage mortality or transplantation	37 (9)	4 (7)	0.97 (0.31–3.04)	0.96	5 (14)	2.12 (0.75–6.00)	0.16
≥1 Unscheduled readmission (any cause)	190 (48)	25 (41)	0.87 (0.49–1.54)	0.63	17 (47)	1.02 (0.51–2.05)	0.95
≥1 Unscheduled readmission for cyanosis	47 (12)	6 (10)	0.91 (0.36–2.31)	0.84	4 (11)	1.02 (0.34–3.05)	0.97

Interstage mortality: 8%  
 No impact of home monitoring on mortality  
 Better weight gain till stage II  
 Effect of general coordinated programme?



Devenir

# Improved Survival of Patients Undergoing Palliation of Hypoplastic Left Heart Syndrome: Lessons Learned From 115 Consecutive Patients

James S. Tweddell, MD; George M. Hoffman, MD; Kathleen A. Mussatto, RN;  
 Raymond T. Fedderly, MD; Stuart Berger, MD; Robert D.B. Jaquiss, MD; Nancy S. Ghanayem, MD;  
 Stephanie J. Frisbee, MSc; S. Bert Litwin, MD

**Early Survival after S1P by Year of Operation**



Figure 1. Hospital survival following stage 1 palliation by year. Hospital survival improved coincident with the application of new treatment strategies beginning in July of 1996 (arrow).

1992-1996: early survival: 53%  
 1996-2001: early survival: 93%

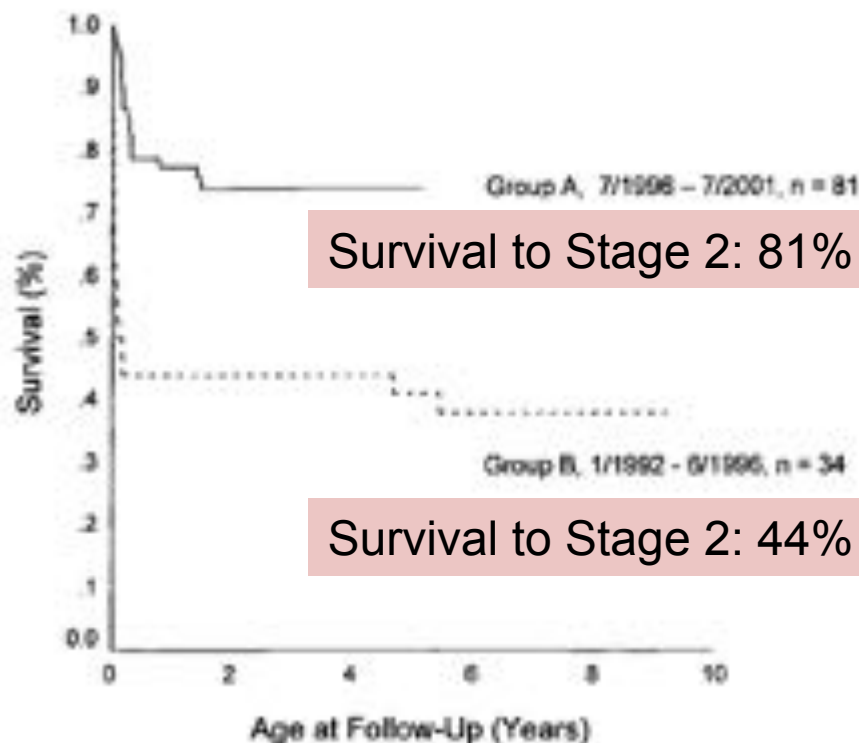


Figure 3. Actuarial survival curve following S1P comparing the time period A (January 1992 to June 1996) with the time period B (July 1996 to October 2001). Survival in the current era is significantly improved with greater than 70% survival to 5 years.



# Expérience de NEM sur 10 ans: HLHS

- 253 fœtus avec DAN HLHS
  - 172 (68%) fœtus: IMG
  - 79 (31%) fœtus: poursuite de grossesse
  - 2 (1%) fœtus: MFIU

*Tableau 17: Atteintes extracardiaques dans l'hypoplasie du cœur gauche.*

	MAJEURE		TOTAL
IMG	23	13 %	172
MFIU	0	0 %	2
NNV	2	2,5 %	79

*MAJEURE= Caryotype anormal et/ou anomalie morphologique majeure.*

# Expérience de NEM sur 10 ans: HLHS

- 79 nouveau-nés vivants
  - 31 (39%) NN: accompagnement
  - 48 (61%) NN: projet chirurgical palliatif

Survie à M12: 11/48 = 23%



# Mortalité en fonction de la CIA

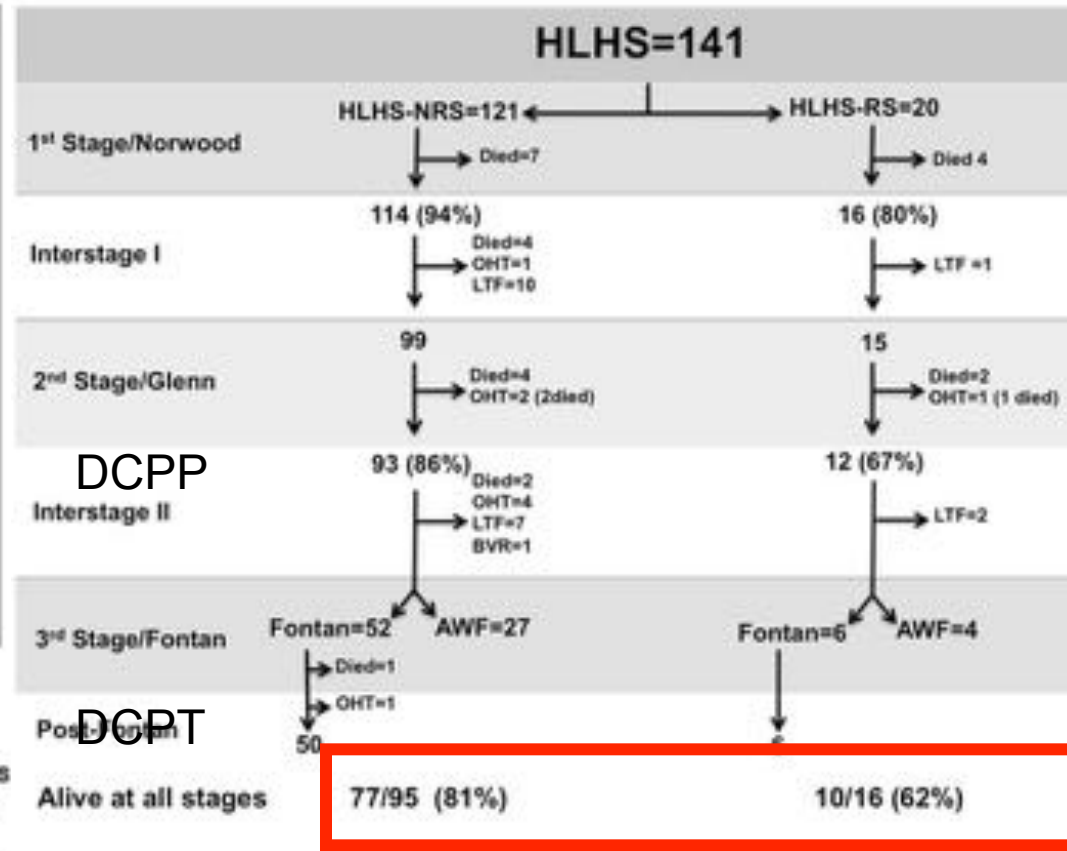
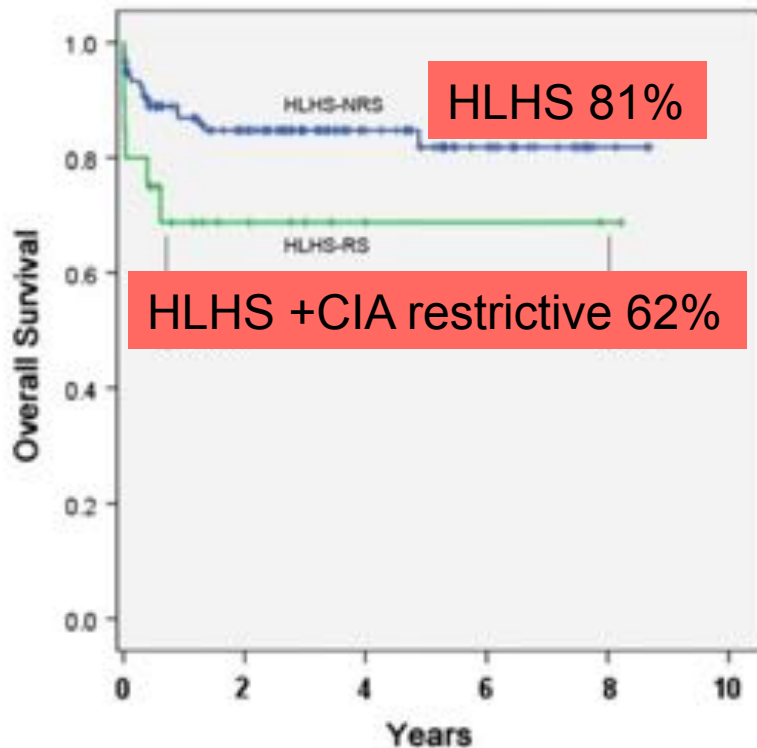
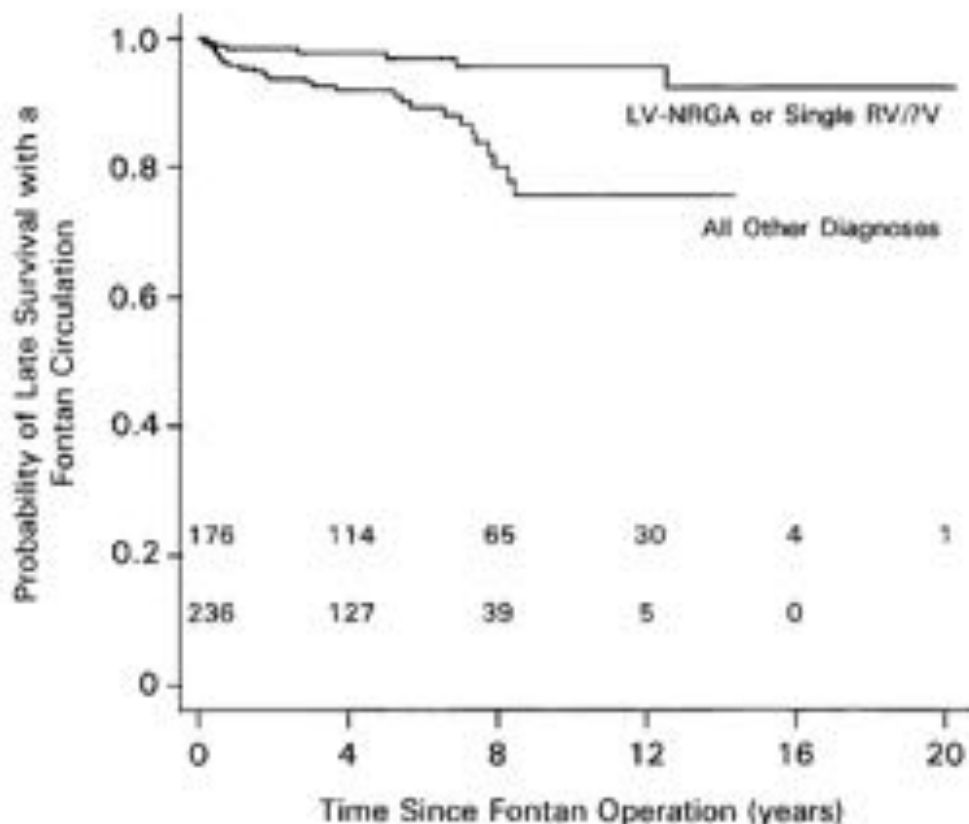


Fig. 3 Comparison of overall survival for HLHS-NRS with HLHS-RS patients

# FONTAN OPERATION IN FIVE HUNDRED CONSECUTIVE PATIENTS: FACTORS INFLUENCING EARLY AND LATE OUTCOME

The Journal of Thoracic and  
Cardiovascular Surgery  
Volume 114, Number 3

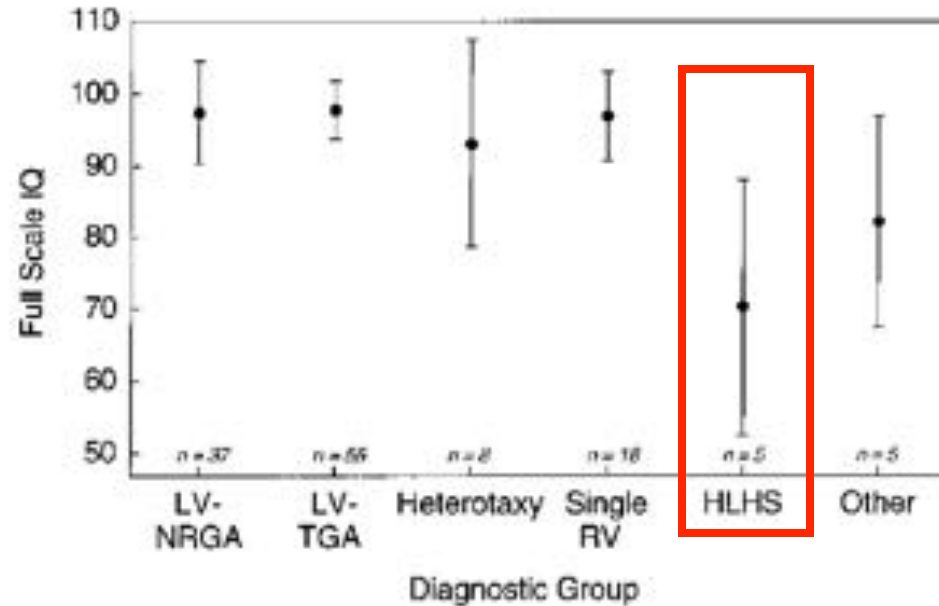
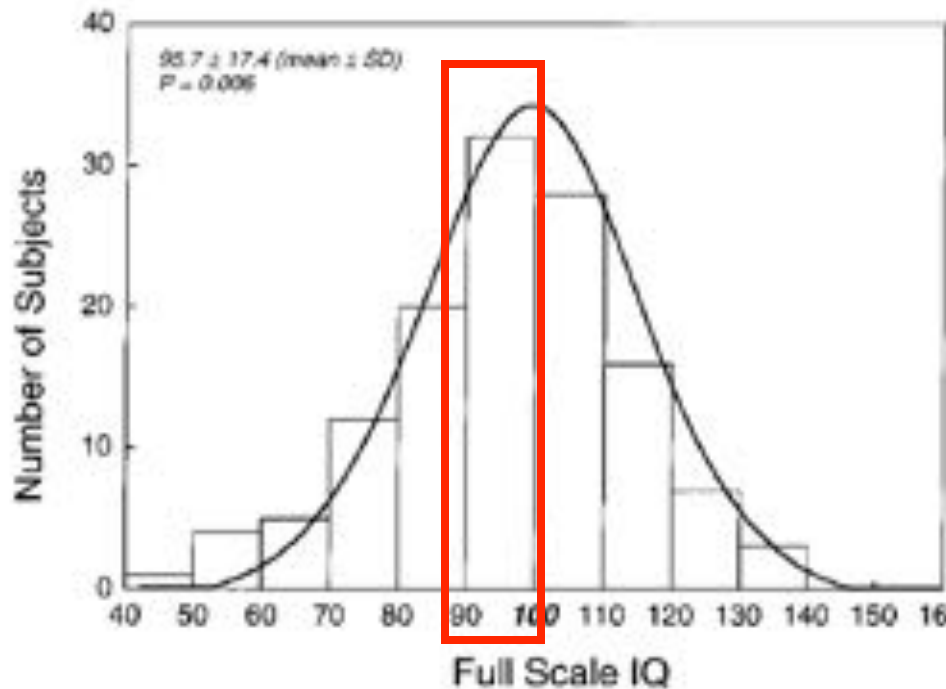


Including HLHS

**Fig. 5.** Kaplan-Meier estimates of the probability of late survival with a Fontan circulation as a function of diagnosis. Time zero is 30 days after the Fontan operation. *LV-NRGA*, Left ventricle with normally related great vessels; *Single RV/?V*, single right ventricle or ventricle with unknown morphology.

# Cognitive Development After the Fontan Operation

Gil Wernovsky, MD; Karen M. Stiles, PhD; Kimberlee Gauvreau, ScD; Thomas L. Gentles, FRACP;



**Mean IQ - ventricule unique: 95**

**Mean-IQ HLHS: 70 ! (n=5)**

# Quality of life and cognitive function in Fontan patients, a population-based study<sup>☆</sup>

Lars Idorn<sup>a,\*</sup>, Annette S. Jensen<sup>a</sup>, Klaus Juul<sup>b</sup>, Dorthe Overgaard<sup>c</sup>, Niels P. Nielsen<sup>d</sup>, Keld Sørensen<sup>e</sup>, Jesper I. Reimers<sup>b</sup>, Lars Søndergaard<sup>a</sup>

**Table 2**  
PedsQL scores by age group in patients and controls.

	5-9 years		p-Value
	Patients (n = 37)	Controls (n = 24)	
Physical health	75 (63-82)	98 (94-100)	<0.001
Psychosocial health	65 (57-72)	92 (87-98)	<0.001
Emotional functioning	70 (50-85)	90 (80-98)	<0.001
Social functioning	70 (60-83)	100 (95-100)	<0.001
School functioning	60 (50-80)	90 (80-100)	<0.001

Data are presented as median (IQR).

	10-15 years		p-Value
	Patients (n = 56)	Controls (n = 34)	
	83 (69-92)	97 (94-100)	<0.001
	80 (66-88)	92 (87-97)	<0.001
	80 (60-85)	90 (75-95)	0.002
	90 (75-98)	100 (95-100)	<0.001
	75 (60-85)	93 (85-100)	<0.001

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International Journal of Cardiology 168 (2013) 3230–3235

**N=10!**

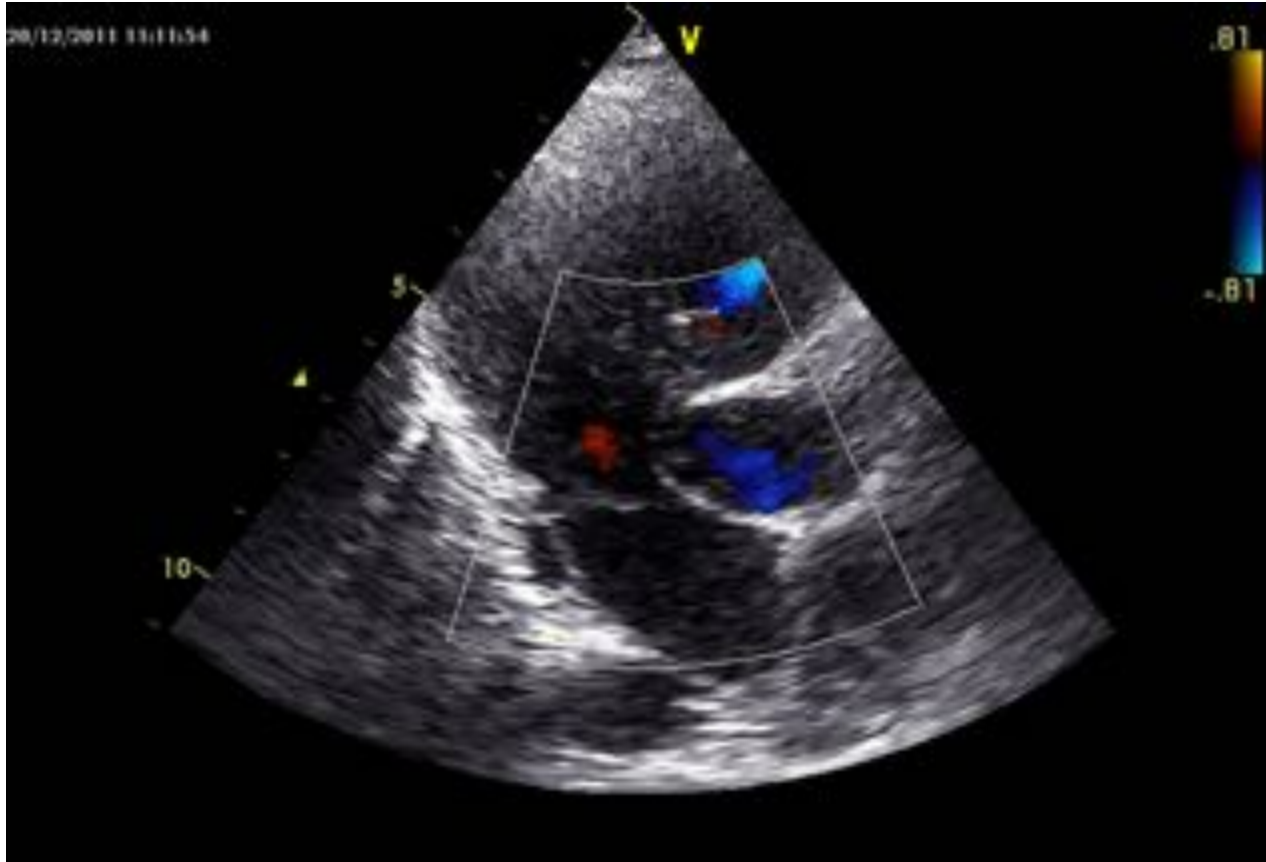
**Table 5**

Comparison of PedsQL and AQF scores between HLHS and non-HLHS patients.

	HLHS	Non-HLHS	p-Value
<i>PedsQL</i>			
<i>Patients</i>			
Physical health, 5–9 years	73.5 (24.1)	72.7 (15.8)	0.94
Physical health, 10–15 years	66.3 (27.6)	80.3 (15.1)	0.07
Psychosocial health, 5–9 years	57.9 (8.1)	66.7 (16.7)	0.31
Psychosocial health, 10–15 years	68.7 (12.8)	76.7 (15.5)	0.27
<i>Parents</i>			
Physical health, 5–9 years	63.3 (20.1)	70.6 (18.5)	0.32
Physical health, 10–15 years	68.4 (20.7)	82.6 (26.9)	0.26
Psychosocial health, 5–9 years	58.1 (10.3)	67.1 (17.8)	0.22
Psychosocial health, 10–15 years	60.0 (11.1)	74.0 (17.2)	0.08
<i>AQF</i>			
Colour, 5–9 years	64.3 (6.3)	48.9 (13.7)	0.07
Colour, 10–15 years	36.2 (9.7)	31.0 (6.7)	0.10
Form, 5–9 years	65.2 (11.9)	56.4 (26.6)	0.35
Form, 10–15 years	33.2 (5.6)	32.4 (9.7)	0.86
Colour-form, 5–9 years	171.0 (33.7)	134.5 (43.4)	0.16
Colour-form, 10–15 years	95.2 (15.6)	73.5 (23.7)	0.05
Overhead, 5–9 years	37.3 (32.0)	29.3 (21.6)	0.57
Overhead, 10–15 years	18.8 (17.4)	10.0 (13.7)	0.19

Data are presented as mean (SD).

# Cas clinique





# Cas clinique

