Interstage programs - single ventricle pathways

Linz approach

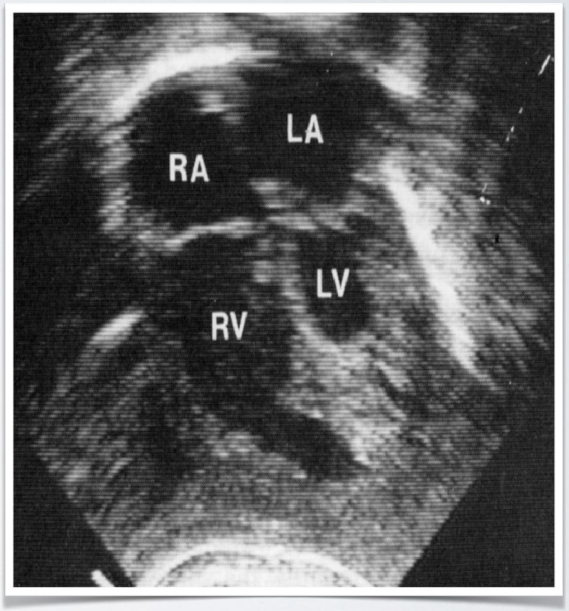
G.Tulzer
Children's Heart Center Linz

Kepler Universitäts Klinikum

Kinder-Herz Zentrum Linz

INTERSTAGE MORTALITY

- Discharge after Norwood to admission for Glenn
- Own data:
  - ✓Interstage mortality in Linz?
  - ✓Risk factors?
- Strategies to prevent interstage mortality
  - ✓Optimize pre-operative status
  - ✓Surgery
  - ✓Medication
  - ✓Monitoring



INTERSTAGE MORTALITY

OWN DATA 1997 - 2022:

- 435 Norwood OPs
  - In-hospital mortality: 11,7% (51/435)
  - Interstage mortality: 6% (23/384)

	N	In-hospital deaths	%	Interstage deaths	%
1997 - 2011	254	40	15,7	18 / 214	8,4
2011 - 2022	181	11	6	5 / 171	2,9

RISK FACTORS

- Prematurity - low birth weight
- Older age at Norwood OP (> 20d)
- Tricuspid valve anomalies
- Anatomy (AA, MS, coronary fistulae…)
- Restrictive / intact atrial septum - TAPVR
- Cardiomyopathies, arrhythmias
- Syndroms - genetic anomalies
- Extracardiac Anomalies …

Linz Approach

European Journal of Cardio-Thoracic Surgery (2014) 1–5
doi:10.1093/ejcts/ezu128

ORIGINAL ARTICLE

Older age at the time of the Norwood procedure is a risk factor for early postoperative mortality†

Eva Sames-Dolzer[a,*], Lale Hakami[a], Michaela Innerhuber[a], Gerald Tulzer[b] and Rudolf Mair[a]

	Group 1 (<20 days)	Group 2 (≥20 days)	*P*-value
N	227	27	
Median age in days [quartiles]	7 [5; 9]	35 [25; 57]	<0.001
Mean weight (kg) ± SD	3.27 ± 0.55	3.71 ± 0.81	0.011
Male sex, *n* (%)	158 (69.6)	19 (70.4)	
Anatomy, *n* (%)			0.002
Aortic atresia	119 (52.4)	4 (14.8)	
Aortic stenosis	66 (29.1)	12 (44.4)	
SV or dominant left ventricle	27 (11.9)	6 (22.2)	
Unbalanced AV canal	15 (6.6)	5 (18.5)	

Kepler Universitäts Klinikum

Kinder-Herz Zentrum Linz

Linz Approach

European Journal of Cardio-Thoracic Surgery (2014) 1–5
doi:10.1093/ejcts/ezu128

ORIGINAL ARTICLE

Older age at the time of the Norwood procedure is a risk factor for early postoperative mortality†

Eva Sames-Dolzer[a,*], Lale Hakami[a], Michaela Innerhuber[a], Gerald Tulzer[b] and Rudolf Mair[a]

	Group 1 (<20 days)	Group 2 (≥20 days)	*P*-value
N	227	27	
Primarily closed chest, *n* (%)	78 (34.4)	4 (14.8)	0.049
NO therapy, *n* (%)	33 (14.5)	11 (40.7)	0.002
Successful CPR, *n* (%)	13 (5.7)	7 (25.9)	0.002
ECMO (since 2008), *n* (%)	7 (9.6)	6 (40)	0.008
30-day mortality, *n* (%)	31 (13.7)	9 (33.3)	0.011
Interstage mortality, *n* (%)	15 (7.7)	3 (16.7)	0.37
Mortality until BDG, *n* (%)	46 (20.3)	12 (44.4)	0.008
Summarized risk for CPR/ECMO/death until BDG, *n* (%)	54 (23.8)	16 (59.3)	<0.001
Median ICU stay (without exitus) in days [quartiles]	12 [9; 16]	11 [9; 17]	0.79
Median hospital stay (without exitus) in days [quartiles]	26 [22; 36]	29 [22; 48]	0.38
Median time between Norwood and BDG in days [quartiles]	115 [99; 129]	100 [85; 125.5]	0.073

Kepler Universitäts Klinikum

Kinder-Herz Zentrum Linz

Unplanned cardiac reoperations and interventions during long-term follow-up after the Norwood procedure†

Eva Sames-Dolzer[a,*], Gregor Gierlinger[a], Michaela Kreuzer[a], Julia Schrempf[a], Roland Gitter[b], Christoph Prandstetter[b], Gerald Tulzer[b], and Rudolf Mair[a]

European Journal of Cardio-Thoracic Surgery 51 (2017) 1044–1050

- 1997 - 2014
- N = 317 Norwood OPs
- Median age: 7d (2 - 270d)
- Median weight: 3,26 kg (24 patients < 2,5 kg)
- BT Shunt: 54, left Sano conduit: 82, right Sano: 181

Unplanned cardiac reoperations and interventions during long-term follow-up after the Norwood procedure†

European Journal of Cardio-Thoracic Surgery 51 (2017) 1044–1050

Mortality reported as a percentage of all patients after Norwood		
Mortality within 30 days after stage I	41/317	12.9
Mortality late interstage I (later than 30 days)	18/317	5.7
Mortality within 30 days after BDG	7/317	2.2
Mortality late interstage II (later than 30 days)	19/317	6.0
Mortality within 30 days after Fontan	1/317	0.3
Mortality late after Fontan (later than 30 days)	5/317	1.6
Total mortality	91/317	28.7
HTX	3/317	0.9
Lost to follow-up	6/317	1.9

Unplanned cardiac reoperations and interventions during long-term follow-up after the Norwood procedure†

European Journal of Cardio-Thoracic Surgery 51 (2017) 1044–1050

Table 2: Unplanned procedures in the interstage I 19%

	No. of patients
Surgical procedure	
Sano conduit change	17
Sano conduit revision	3
mBTS instead of Sano conduit	2
mBTS change	6
Tricuspid valve repair	3
Aortic arch repair	2
Atrial thrombectomy	1
Pacemaker implantation	1
Total	35
Interventional procedure	
Aortic arch dilatation	11
RV-PAC stent (1 plus isthmus stent)	7
APCA coilings	5
LSVC coiling	1
LPA dilatation	1
Total	25

Unplanned cardiac reoperations and interventions during long-term follow-up after the Norwood procedure†

European Journal of Cardio-Thoracic Surgery 51 (2017) 1044–1050

Table 3: Unplanned procedures in the interstage II

	No. of patients
Surgical procedure	
Tricuspid valve repair	6
Additional azygos vein ligation	1
Ligation of major veno-venous collaterals	1
Pacemaker implantation	1
Total	9
Interventional procedure	
Coiling of aorto-pulmonary collaterals (1 + LPA dilatation)	17
Stenting of LPA stenosis	6
Dilatation of AA restenosis (1 + Rashkind)	4
Stenting of AA restenosis (2 + LPA stent)	4
Coiling of LSVC	2
Coiling of veno-venous fistulas	3
Coiling of pulmonary AV-fistulas	2
Closure of native aortic valve	1
Total	39

Linz Approach

Unplanned cardiac reoperations and interventions during long-term follow-up after the Norwood procedure†

Eva Sames-Dolzer[a,*], Gregor Gierlinger[a], Michaela Kreuzer[a], Julia Schrempf[a], Roland Gitter[b], Christoph Prandstetter[b], Gerald Tulzer[b], and Rudolf Mair[a]

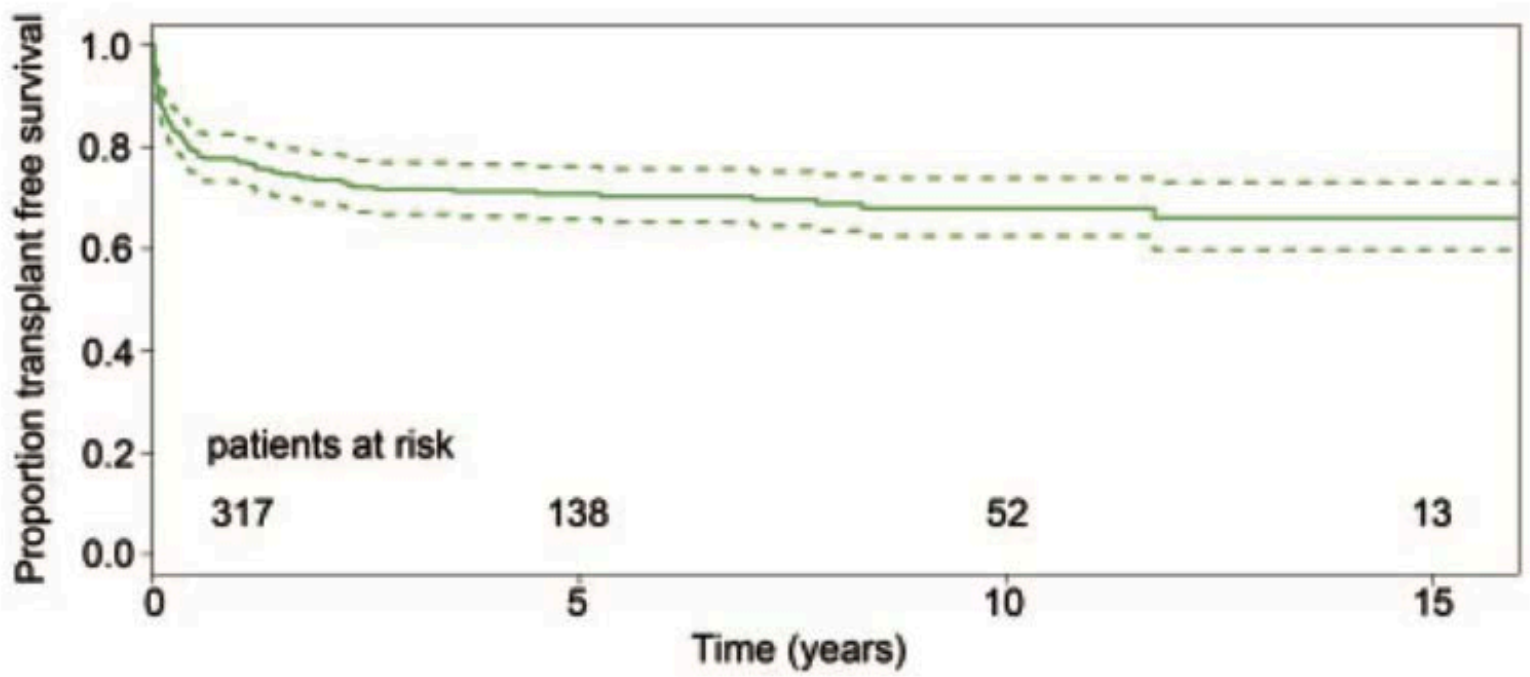


Figure 1: Transplant free survival for the whole Norwood cohort.

Kepler Universitäts Klinikum

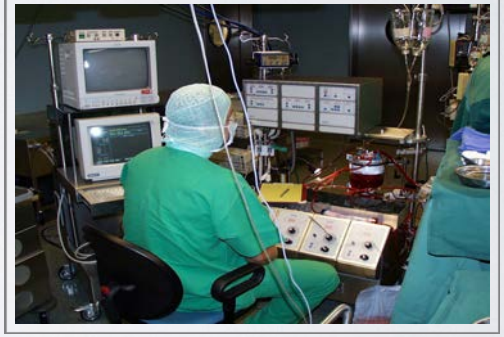
Kinder-Herz Zentrum Linz

REDUCE RISK FACTORS

- Prematurity - low birth weight ➡Delivery at term
- Older age at Norwood OP (> 20d) ➡Norwood before 20d or hybrid after 20d?
- Tricuspid valve anomalies ➡???
- Anatomy (AA, MS, coronary fistulae…) ➡???
- Restrictive / intact atrial septum - TAPVR ➡Prenatal intervention/ PAB pre Norwood
- Cardiomyopathies, arrhythmias ➡CHF therapy
- Syndroms - genetic anomalies ➡???
- Extracardiac Anomalies … ➡???

SURGERY

- Right time
- Sano conduit instead of mBT Shunt
- Address TV problems
- Address impaired pulm. venous drainage
- Aortic arch reconstruction *
- New perfusion techniques **



*Sames-Dolzer E. et al: Aortic arch reconstruction in the Norwood procedure using a curved polytetrafluoroethylene patch. Eur J Cardiothorac Surg 2022

** Kreuzer M et al: double arterial cannulation during aortic arch reconstruction in pediatric patients. Multimed Man Cardiothorac Surg 2018

MEDICATION

- Balance Qp/Qs (Rp/Rs) - pulmonary vasodilators
- Manage CHF (milrinone, levosimendan, diuretics, ACE inhibitors, beta-blockers, digoxin)
- Manage arrhythmias

MONITORING

- Train the parents
- HR monitor if arrhythmias
- Frequent ambulatory visits

and:

- Nutrition!
- Early Glenn at 3-4 months

Interstage programs - single ventricle pathways

Linz approach

Summary

Each patient requires an individual treatment according to

- Age
- Anatomy
- Risk factors
- Postoperative course

Kepler Universitäts Klinikum

Kinder-Herz Zentrum Linz