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Centro de Referencia Perinatal Oriente
Facultad de Medicina, Universidad de Chile

Sindrome Corazón Izquierdo Hipoplasico : Factores de riesgos de mortalidad

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Introducción

El término síndrome del corazón izquierdo hipoplásico (SCIH) fue descrito por Noonan y Nadas en 1958.

Definición: Subdesarrollo grave de lado izquierdo del corazón, incapaz de realizar la circulación sistémica.

ANATOMIA + FUNCIONAL DEL LADO IZQUIERDO DEL CORAZÓN

Generalidades

El HLHS tiene una prevalencia al nacer de aproximadamente 2 a 3 casos por cada 10 000 nacidos vivos en los Estados Unidos

Representa del 1 al 3% de todas las cardiopatías congénitas .

Se observa un predominio masculino (proporción hombre:mujer de aproximadamente 1,5:1).

10% Asociado aneuploideas: Trisomia 13 -18 -21

25% con genopatias: 40% Sd Turner

Etiología

Obstrucción en el TSVI
Estenosis aortica severa/AO

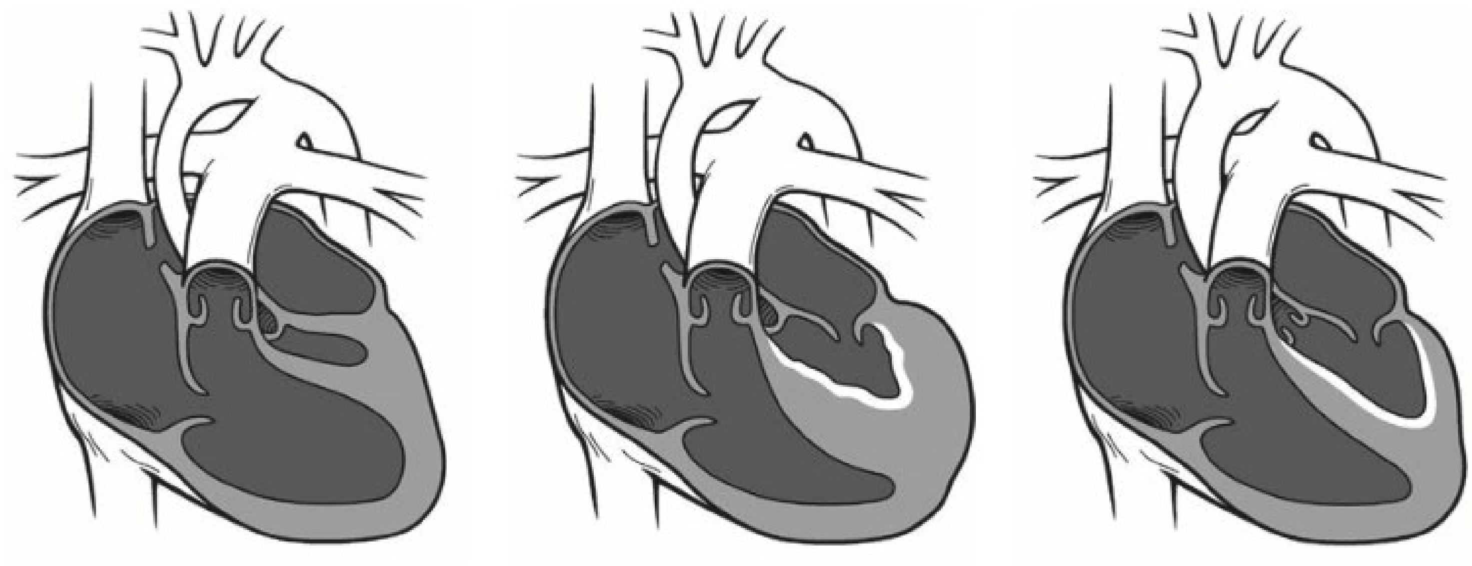
Disminución del flujo de entrada en el VI
Estenosis mitral – Alteración en el septum interucricular

Anomalía intrenseca del desarrollo del miocardio del VI

Disminución del flujo sanguineo transventricular

Desarrollo deficiente del VI

Fenotipos

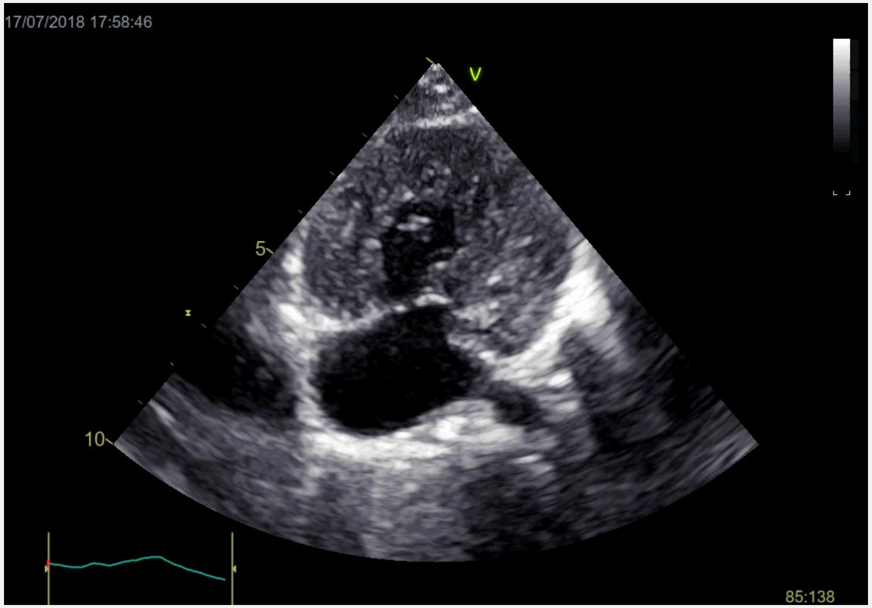


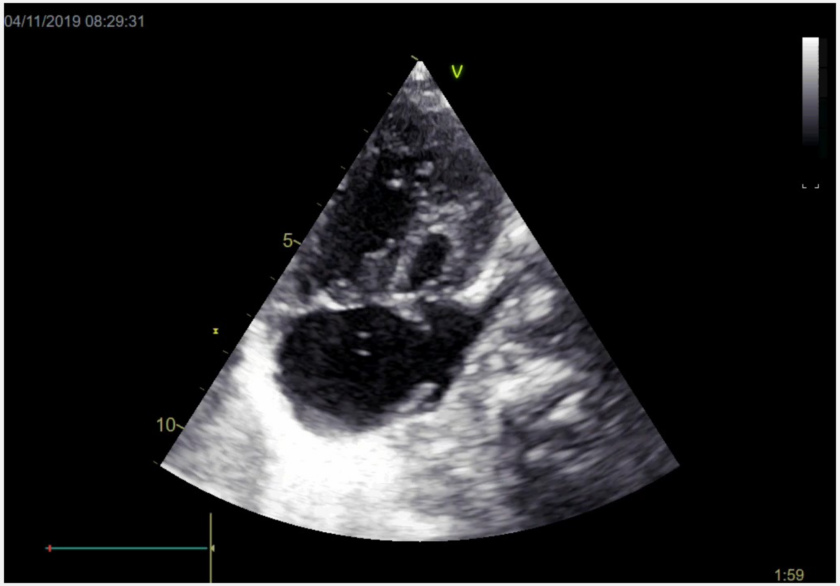
A Aortic valve atresia with mitral valve atresia

B Aortic valve atresia with patent mitral valve

C Aortic valve stenosis with patent mitral valve

Fenotipos





Tratamiento

Manejo conservado

1983……
Actualidad

Cx palitiava

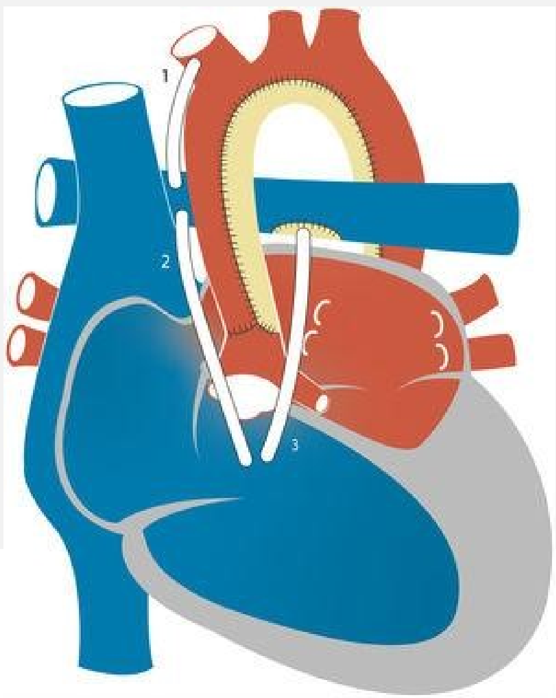
Transplante

Sin indicación Qx

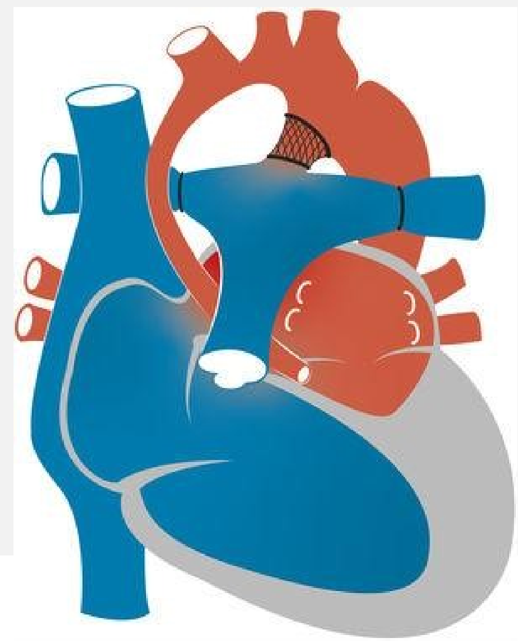
Tratamiento

Objetivos:

- Flujo de salida sistémico sin obstrucciones,
- Retorno venoso pulmonar sin restricciones
- Flujo sanguíneo pulmonar restringido



Cx norwoood



Procedimiento
Hibrido

SCIH:

- Es una de las formas más graves de cardiopatía congénita
- Una de las más difíciles de tratar
- Aún hoy, uno de los más discutidos y debatidos de enfermedades cardiovasculares congénitas

Cx Norwood: la supervivencia de los recién nacidos con SCIH en la infancia ha mejorado con cada.

Mejoría de la técnica quirúrgica

Diagnóstico prenatal

Cuidado preoperatorio y anestésico

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Circulation
Volume 92, Issue 9, 1 November 1995; Pages 262-266
https://doi.org/10.1161/01.CIR.92.9.262

American Heart Association.

ARTICLE

Ten-Year Institutional Experience With Palliative Surgery for Hypoplastic Left Heart Syndrome

Risk Factors Related to Stage I Mortality

Joseph M. Forbess, Nancy Cook, Stephen J. Roth, Alain Serraf, John E. Mayer, Jr, and

Outcome of the Norwood operation in patients with hypoplastic left heart syndrome: A 12-year single-center survey

Article in The Journal of thoracic and cardiovascular surgery · October 2009

CONGENITAL: NORWOOD

Thirty years and 1663 consecutive Norwood procedures: Has survival plateaued?

Check for updates

Christopher E. Mascio, MD,[a] Mallory L. Irons, MD, MBE,[b] Richard F. Ittenbach, PhD,[c] J. William Gaynor, MD,[a] Stephanie M. Fuller, MD,[a] Michelle Kaplinski, MD,[d] Andrea T. Kennedy, BS,[e] James M. Steven, MD,[f] Susan C. Nicolson, MD,[f] and Thomas L. Spray, MD[a]

patients indicate improved survival for patients with hypoplastic left heart syndrome (HLHS) undergoing stage I palliation with a right ventricle to pulmonary artery conduit (NW-RVPA) vs. a modified Blalock-Taussig shunt (NW-BT). We compared postoperative outcomes in a concurrent series of patients with HLHS undergoing an NW-BT procedure vs. NW-RVPA procedure.

Design: Perioperative data from 66 consecutive patients who underwent NW-BT ($n = 37$) or NW-RVPA ($n = 29$) procedures were retrospectively analyzed.

Setting: Cardiac intensive care unit in a tertiary pediatric hospital.

Patients: Charts were reviewed for all patients with the diagnosis of HLHS undergoing the NW-BT or NW-RVPA procedure between January 2002 and December 2003.

Results: Cardiopulmonary bypass time was longer in the NW-BT group than in the NW-RVPA group (152.5 ± 52.0 vs. 134.5 ± 36.1 mins; $p = .04$). Postoperative diastolic pressures were

NW-RVPA group over the first 72 hrs. Time to sternal closure (2 [1–6] vs. 4 [2–41] days; $p = .01$), duration of mechanical ventilation (113 [49–386] vs. 136 [84–764] hrs; $p = .01$), time to establish enteral feeds (4 [2–8] vs. 5 [3–22] days; $p = .01$), length of intensive care unit stay (11 [7–55] vs. 15 [8–90] days; $p = .04$), and length of hospital stay (16 [11–67] vs. 27 [12–126] days; $p = .01$) were shorter in the NW-RVPA group. Postoperative mortality was not significantly different between the NW-RVPA group (7%) and NW-BT group (11%).

Conclusion: At an experienced institution with low stage I palliation mortality for HLHS, there were no differences in early morbidity and mortality between the NW-RVPA and NW-BT procedures. The primary advantage of the NW-RVPA procedure may be faster recovery following surgery and earlier discharge from the hospital. (Pediatr Crit Care Med 2006; 7:238–244)

Key Words: congenital heart disease; Norwood operation; hypoplastic left heart syndrome

ised mortality compared to ventricular ts without antegrade mitral flow. All S born between 2002 and 2011 year who Norwood palliation with complete history ıms for patency of the mitral and aortic ed. The cohort was divided into three sia–mitral atresia (AA–MA), aortic ste- sis (AS–MS), and aortic atresia–mitral . Demographic, comorbidities, and oper-

The Norwood procedure for mitral stenosis and aortic atresia is more often associated with use of extracorporeal membrane oxygenation and shunt revision compared to other variants. This HLHS variant has lowest long-term survival.

Keywords Hypoplastic left heart syndrome · Norwood procedure · Outcomes

CONGENITAL: NORWOOD

Thirty years and 1663 consecutive Norwood procedures: Has survival plateaued?

Check for updates

Christopher E. Mascio, MD,[a] Mallory L. Irons, MD, MBE,[b] Richard F. Ittenbach, PhD,[c] J. William Gaynor, MD,[a] Stephanie M. Fuller, MD,[a] Michelle Kaplinski, MD,[d] Andrea T. Kennedy, BS,[e] James M. Steven, MD,[f] Susan C. Nicolson, MD,[f] and Thomas L. Spray, MD[a]

The Journal of Thoracic and Cardiovascular Surgery • July 2019

Estudio restrospectivo 1 de enero de 1984 al 6 de mayo de 2014.
Se incluyeron todos los sujetos que se sometieron a un procedimiento de Norwood entre estas fechas (HLHS y todas las variantes).
No hubo criterios de exclusión

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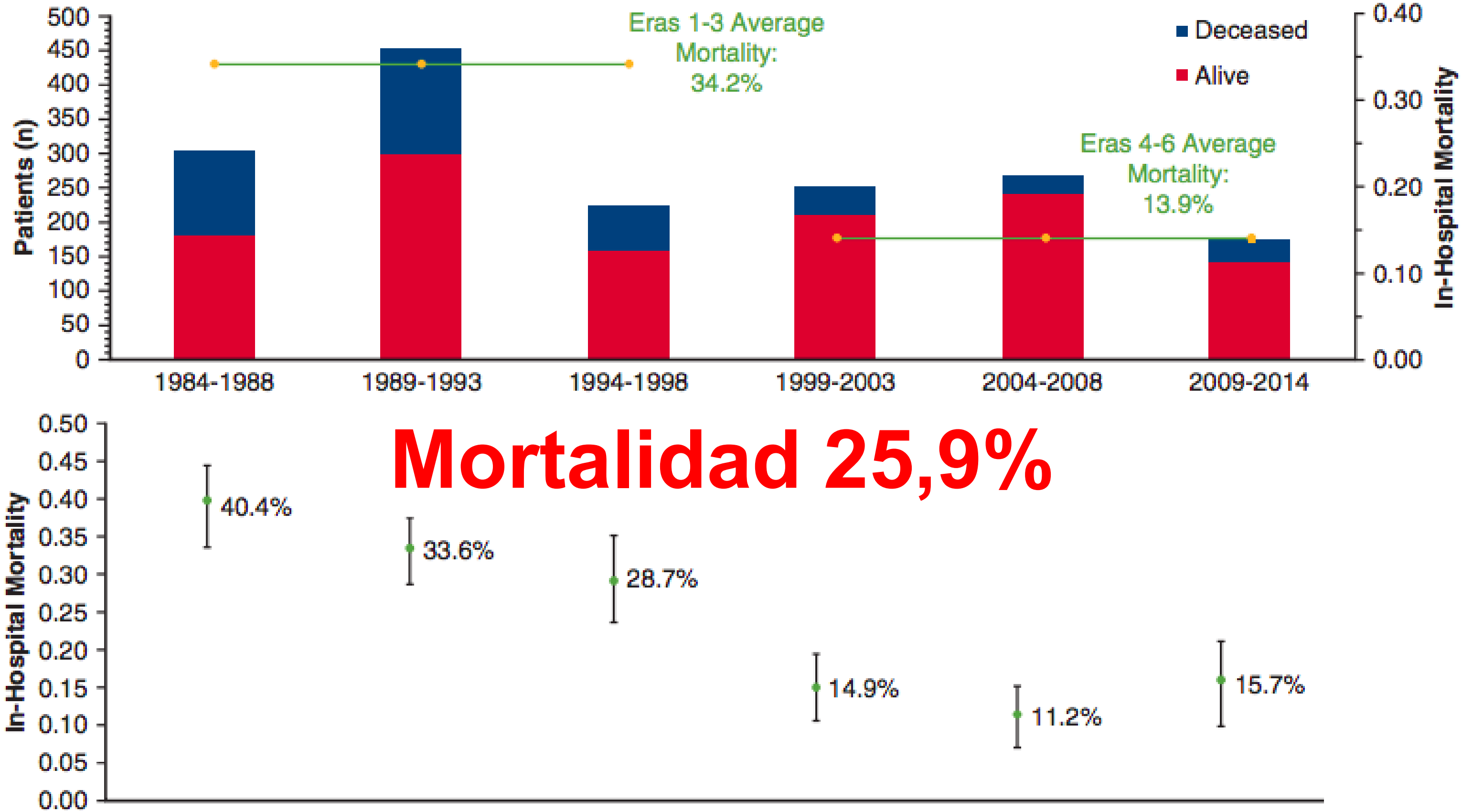
Objetivos:

Analizar la mortalidad hospitalaria a lo largo del tiempo

Describir los factores de riesgo específicos del paciente en la mortalidad.

Era	
1984-1988	302 (18.2)
1989-1993	450 (27.1)
1994-1998	223 (13.4)
1999-2003	249 (15.0)
2004-2008	267 (16.1)
2009-2014	172 (10.3)



Mortalidad 25,9%

FIGURE 1. Hospital mortality has decreased after the Norwood procedure compared with earlier eras.

TABLE 3. Frequency and mortality of anatomic subtypes undergoing the Norwood procedure

Anatomy	Frequency	Mortality
HLHS variant	34.8% (519/1492)	26.8%
Aortic atresia/mitral atresia	32.2% (480/1492)	26.7%
Aortic atresia/mitral stenosis	16.4% (244/1492)	27.0%
Aortic stenosis/mitral stenosis	15.4% (230/1492)	20.4%
Aortic stenosis/mitral atresia	1.27% (19/1492)	31.6%

HLHS, Hypoplastic left heart syndrome.

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TABLE 1. Patient characteristics (N = 1663)

Variable	Frequency		n	Median
	No (%)	Yes (%)		(IQ1, IQ3)
Anomalous pulmonary venous drainage	1491 (89.7)	81 (4.9)		
Atrioventricular valve regurgitation	1450 (87.2)	181 (10.9)		
Intact or highly restrictive atrial septum	1379 (82.9)	243 (14.6)		
Preterm birth (<37 wk)	1312 (78.9)	205 (12.3)		
Era				
1984-1988		302 (18.2)		
1989-1993		450 (27.1)		
1994-1998		223 (13.4)		
1999-2003		249 (15.0)		
2004-2008		267 (16.1)		
2009-2014		172 (10.3)		
Gender				
Female		644 (38.7)		
Male		1019 (61.3)		
Genetic anomaly				
Normal		1128 (67.8)		
Suspect/abnormal		165 (9.9)		
HLHS subtype				
Aortic atresia/mitral atresia		480 (28.9)		
Aortic atresia/mitral stenosis		244 (14.7)		
Aortic stenosis/mitral atresia				
Atrial stenosis/mitral stenosis		249 (15.0)		
HLHS variant		519 (31.2)		
Race				
White		1154 (69.4)		
African-American		258 (15.5)		
Other		245 (14.7)		
Ascending aorta (mm)			1266	3.0 (2.0, 4.6)
Age at Norwood			1663	5.3 (3.3, 8.3)
Birth weight (g)			1529	3.2 (2.8, 3.5)
Weight at Norwood			1630	3.2 (2.8, 3.6)

IQ, Interquartile; *HLHS*, hypoplastic left heart syndrome.

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TABLE 6. Mortality by total number of risk factors and era (N = 1663)

Era (mortality, y)	No. of identified risk factors per subject: 0	1	2	≥3
Era 1 (40.4%, 1984-1988)	59 (39.3)	18 (47.4)	18 (30.0)	22 (46.8)
Era 2 (33.6%, 1989-1993)	63 (29.0)	23 (36.5)	32 (30.5)	33 (50.8)
Era 3 (28.7%, 1994-1998)	27 (25.2)	6 (24.0)	15 (31.2)	16 (37.2)
Era 4 (14.9%, 1999-2003)	5 (8.2)	5 (12.8)	9 (11.4)	18 (25.7)
Era 5 (11.2%, 2004-2008)	5 (12.2)	3 (4.8)	8 (9.8)	14 (17.3)
Era 6 (15.7%, 2009-2014)	1 (6.2)	7 (12.5)	5 (10.6)	14 (26.4)
Total (25.9%; 1984-2014)	160 (27.0)	62 (21.8)	87 (20.8)	117 (32.6)

For purposes of this analysis, risk factors were presence of anomalous pulmonary venous drainage, moderate to severe atrioventricular valve regurgitation, confirmed/suspected genetic anomaly, intact/highly restrictive atrial septum, preterm birth (dichotomized at 37 weeks), and weight at stage 1 reconstruction (dichotomized at 2.5 kg).

FACTORES DE RIESGO MORTALIDAD

- ✓ GENOPATIA
- ✓ PESO <2,5 kg
- ✓ PREMATUREZ

SEPTUM INTACTO

DRENAJE VENOSO ANOMALO PULMONAR TOTAL

DISFUNCION DEL VD

INSUFICIENCIA TRICUSPIDEA

AORTA ASCENDENTE DIMINUTA

SCIH/SEPTUM INTACTO

- el 6 % de SCIH
- continúa teniendo tasas de mortalidad superior al 50%.

Cambios vasculares pulmonares durante la gestación como resultado de una aurícula izquierda hipertensa.

marcada linfangiectasia e hipoplasia pulmonar que sugiere lesión pulmonar en etapa terminal.

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Cardiology in the Young (2016), 26, 1072–1081
doi:10.1017/S1047951115001791

Original Article

Management of hypoplastic left heart syndrome with intact atrial septum: a two-centre experience

Shyam K. Sathanandam,[1,2] Ranjit Philip,[2] David Gamboa,[1,3] Andrew Van Bergen,[1] Michel N. Ilbawi,[1] Christopher Knott-Craig,[2] B. Rush Waller,[2] Alexander J. Javois,[1] Bettina F. Cuneo[1,4]

[1]*Advocate Children's Hospital, Oak Lawn, Illinois;* [2]*University of Tennessee Health Science Center, LeBonheur Children's Hospital Memphis, Tennessee;* [3]*School of Medicine, The University of Utah, Salt Lake City, Utah;* [4]*Children's Hospital Colorado, Aurora, Colorado, United States of America*

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Table 1. Comparison of variables between the three subgroups.

Variable Total = 158	Group-1 n = 140	Group-2 n = 11	Group-3 n = 7	p-Value*
Pre-operative variables				
Gender (male/female)	87/53	8/3	6/1	0.5
Birth weight (kg)	3.2 ± 0.5	3.3 ± 0.4	3.0 ± 0.4	0.8
Prenatal diagnosis	112 (80%)	11 (100%)	0%	<0.05
Foetal PVD VTIR/VTIF%	13% (5–18%)	49% (24–72%)	N/A	<0.001
Diagnosis				<0.001
MS-AA	36 (26%)	1 (9%)	2 (29%)	
MA-AA	45 (32%)	8 (73%)	4 (57%)	
MS-AS	59 (42%)	2 (18%)	1 (14%)	
Size of AAO (mm)	3.4 ± 1.6	2.6 ± 0.4	2.6 ± 0.7	0.01
Operative variables				
Age at S1P (days)	4.5 ± 2.1	5.4 ± 2.8	9.8 ± 7.4	0.5
Type of S1P				<0.001
Hybrid	12 (8%)	6 (55%)	4 (57%)	
Sano	96 (68%)	2 (18%)	3 (43%)	
Shunt	32 (24%)	1 (9%)	0 (0%)	
Transplant before S1P	0 (0%)	2 (18%)	0 (0%)	
Bypass time (minutes)	106.4 ± 22.5	100.2 ± 21.8	108.0 ± 31.6	0.1
Cross-clamp time (minutes)	48.6 ± 16.3	46.4 ± 19.3	50.8 ± 23.6	0.5
Circulatory arrest time (minutes)	40.8 ± 9.3	40.6 ± 9.2	44.3 ± 16.4	0.2
Antegrade perfusion time (minutes)	35.5 ± 7.2	36.7 ± 8.6	37.9 ± 10.8	0.3
Post-operative variables				
ECMO	17 (12%)	3 (27%)	4 (57%)	<0.05
Hospital LOS (days)	24.4 ± 16.2	32.1 ± 21.7	34.5 ± 24.6	0.4
ICU LOS (days)	20.6 ± 15.4	26.4 ± 15.8	31.4 ± 21.9	0.5
Survival to S2P	116 (83%)	5 (46%)	0	<0.001
Heart Transplantation	3 (2%)	2 (18%)	0	<0.05
Completed Fontan/awaiting Fontan	96/20	4/1	0	–

AAO = ascending aorta; ICU = Intensive Care Unit; LOS = length of stay; S1P = stage 1 palliation; S2P = stage 2 palliation

*Comparing Groups 1 and 2

DVPAT

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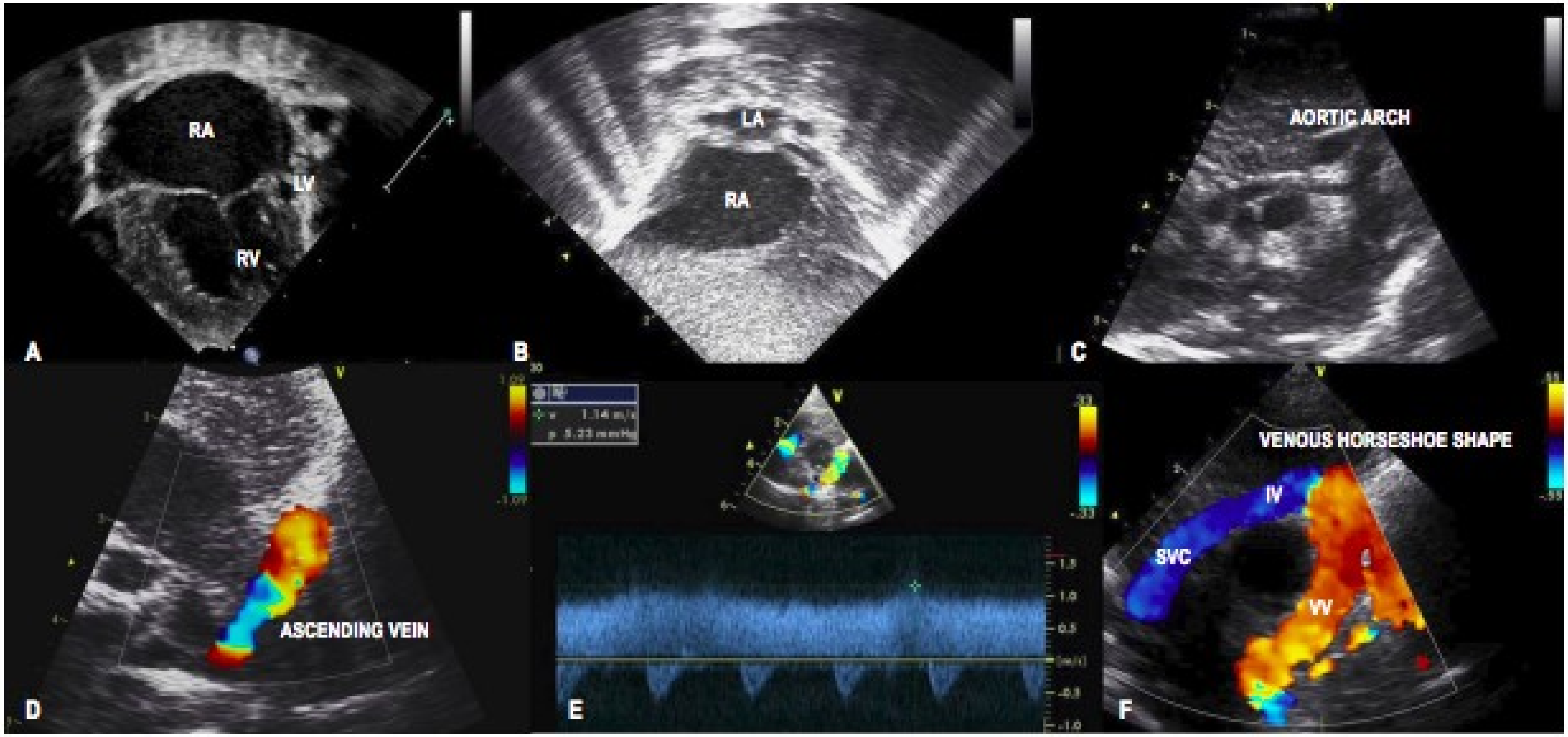
- la incidencia de complicaciones de la conexión venosa pulmonar anómala total (TAPVC) en pacientes con síndrome del corazón izquierdo hipoplásico es baja (2,3%)
- el pronóstico es malo.


RA
LV
RV
A
LA
RA
B
AORTIC ARCH
C
ASCENDING VEIN
D
E
VENOUS HORSESHOE SHAPE
IV
SVC
VV
F

FACTORES DE RIESGO MORTALIDAD

- ✓ GENOPATIA
- ✓ PESO <2,5 kg
- ✓ PREMATUREZ

- ✓ SEPTUM INTACTO
- ✓ DRENAJE VENOSO ANOMALO PULMONAR TOTAL

DISFUNCION DEL VD

INSUFICIENCIA TRICUSPIDEA

AORTA ASCENDENTE DIMINUTA

Insuficiencia Triscúspidea

- IT se observa en el 25% de los niños con síndrome del corazón izquierdo hipoplásico.
- 10% IT significativa
- IT significativa se asocia con una peor supervivencia a 1 año y la reparación exitosa puede resultar en una supervivencia equivalente a 5 años.
- En aquellos con IT persistente y disfunción del ventrículo derecho, sin embargo, la mortalidad permaneció alta en 80%

Insuficiencia Triscúspidea

Reparación de la válvula tricúspide en los recién nacidos sometidos a la operación de Norwood no se realiza en esta etapa debido al estado fisiológico frágil y anatómicamente complejas de la válvula tricúspide y, por lo tanto, se ha limitado a la reparación posterior antes, durante o después de la derivación de Glenn bidireccional.

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European Journal of Cardio-Thoracic Surgery 54 (2018) 993–1000
doi:10.1093/ejcts/ezy198 Advance Access publication 19 May 2018

ORIGINAL ARTICLE

Cite this article as: Alsoufi B, Sinha R, McCracken C, Figueroa J, Altin F, Kanter K. Outcomes and risk factors associated with tricuspid valve repair in children with hypoplastic left heart syndrome. Eur J Cardiothorac Surg 2018;54:993–1000.

Outcomes and risk factors associated with tricuspid valve repair in children with hypoplastic left heart syndrome†

Bahaaldin Alsoufi*, Raina Sinha, Courtney McCracken, Janet Figueroa, Firat Altin and Kirk Kanter

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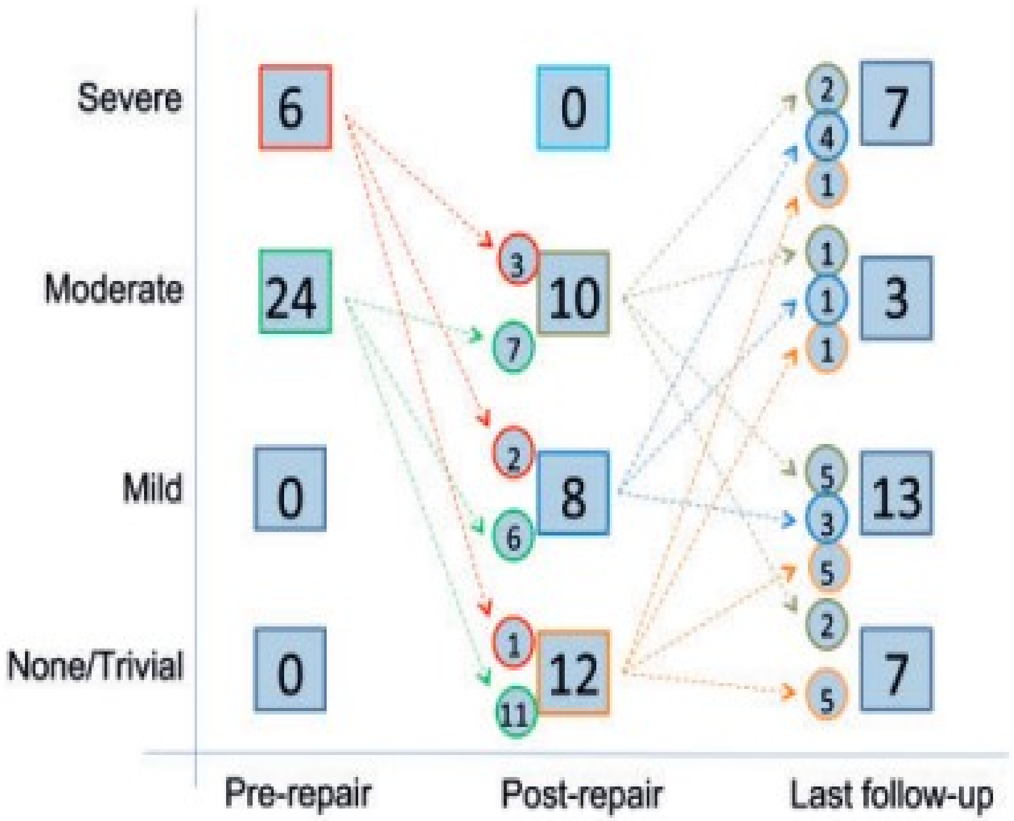
Revisamos los registros de todos los neonatos con SCIH (n = 219) que se sometió a la operación de Norwood en Children's Healthcare de Atlanta, Emory University de 2002 a 2012.

30 pctes requirieron reparación de la VT

RESULTADOS



Severe
Moderate
Mild
None/Trivial
6
24
0
0
0
10
8
12
3
7
2
6
1
11
7
3
13
7
2
4
1
1
1
1
5
3
5
2
5
Pre-repair
Post-repair
Last follow-up

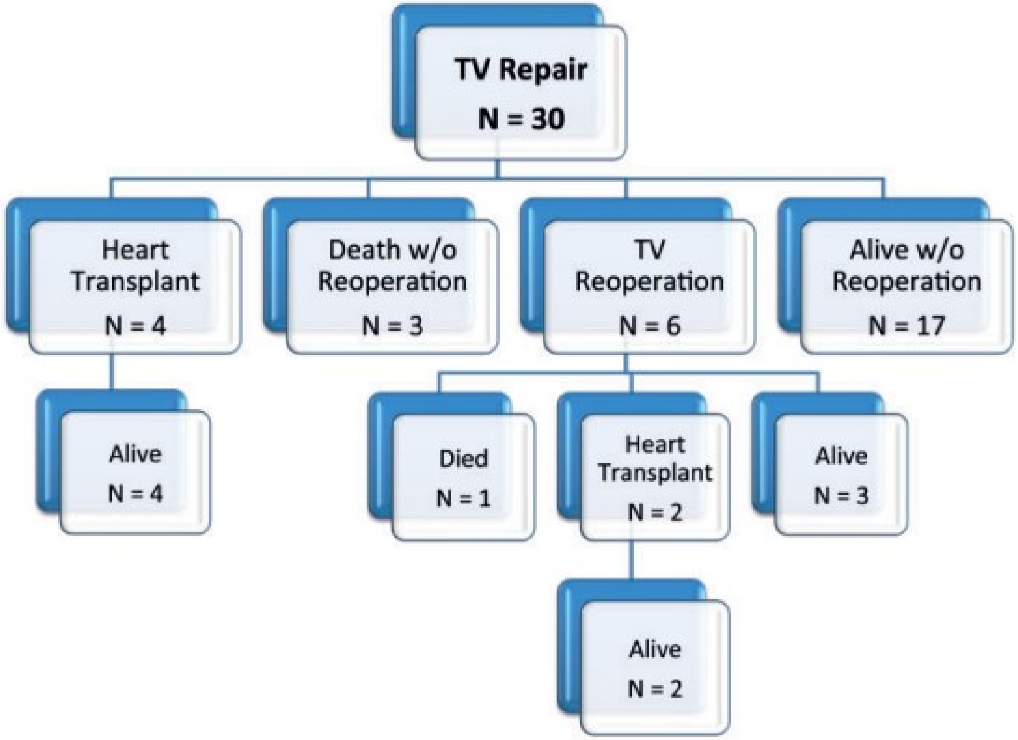


Figure 3: A flowchart showing the clinical progress following TV repair in 30 patients with hypoplastic left heart syndrome. TV: tricuspid valve.

El análisis de riesgo competitivo mostró que 10 años después de la reparación de TV:

- el 21 % de los pacientes se sometieron a una nueva operación de TV
- el 18 % murió o fueron trasplantados
- el 61% estaban vivos sin reintervención posterior.

FACTORES DE RIESGO MORTALIDAD

- ✓ GENOPATIA
- ✓ PESO <2,5 kg
- ✓ PREMATUREZ

- ✓ SEPTUM INTACTO
- ✓ DRENAJE VENOSO ANOMALO PULMONAR TOTAL
- ❖ DISFUNCION DEL VD
- ❖ INSUFICIENCIA TRICUSPIDEA

AORTA ASCENDENTE DIMINUTA

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Does Ascending Aorta Size Affect Norwood Outcomes in Hypoplastic Left Heart With Aortic Atresia?

Check for updates

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Division of Cardiothoracic Surgery, Department of Surgery, Washington University School of Medicine, St Louis, Missouri; and Section of Pediatric Cardiothoracic Surgery, St Louis Children's Hospital, St Louis, Missouri

293 pacientes

Table 2. Overall Outcomes for Aorta Size Groups

Outcome	Diminutive ≤1.5 mm (n = 29)	Small 1.6-1.9 mm (n = 57)	Medium 2.0-3.9 mm (n = 181)	Large ≥4.0 mm (n = 25)	*P* Value
Early Outcomes					
14-day mortality	6 (20.7)	4 (7.0)	7 (3.9)	1 (4.0)	.006
Transplant/death at 30 d	8 (27.6)	5 (8.8)	16 (8.8)	1 (4.0)	.012
ECMO at surgery	4 (13.8)	3 (5.3)	13 (7.2)	1 (4.0)	.459
ECMO/CPR during hospitalization	8 (27.6)	12 (21.1)	30 (16.6)	3 (12.0)	.392
Duration of mechanical ventilation (d)	8 (5-12.5)	6 (4.3-14)	7 (5-12)	7 (4-13.5)	.681
ICU-free days to 28 d	0 (0-12)	8 (0-17)	13 (0-18)	9 (0-19)	.009
Late Outcomes					
Transplant/death at 14 mo	13 (44.8)	13 (22.8)	57 (31.5)	4 (16.0)	.072
RV fractional area change (%)[a]	29 ± 8	34 ± 7	32 ± 8	35 ± 4	.487
RV ejection fraction (%)[b]	42.7 ± 6.1	42.8 ± 10.3	42.9 ± 8.5	45.1 ± 5.4	.856
RV end diastolic volume (mL)[b]	38.4 ± 12.7	32.6 ± 8.4	32.8 ± 12.2	30.9 ± 5.9	.391
RV end systolic volume (mL)[b]	22.1 ± 8.0	18.8 ± 6.1	19.2 ± 10.3	17.0 ± 3.9	.614
Significant TR (≥moderate)[c]	6 (42.9)	6 (17.1)	30 (25.9)	5 (31.3)	.270

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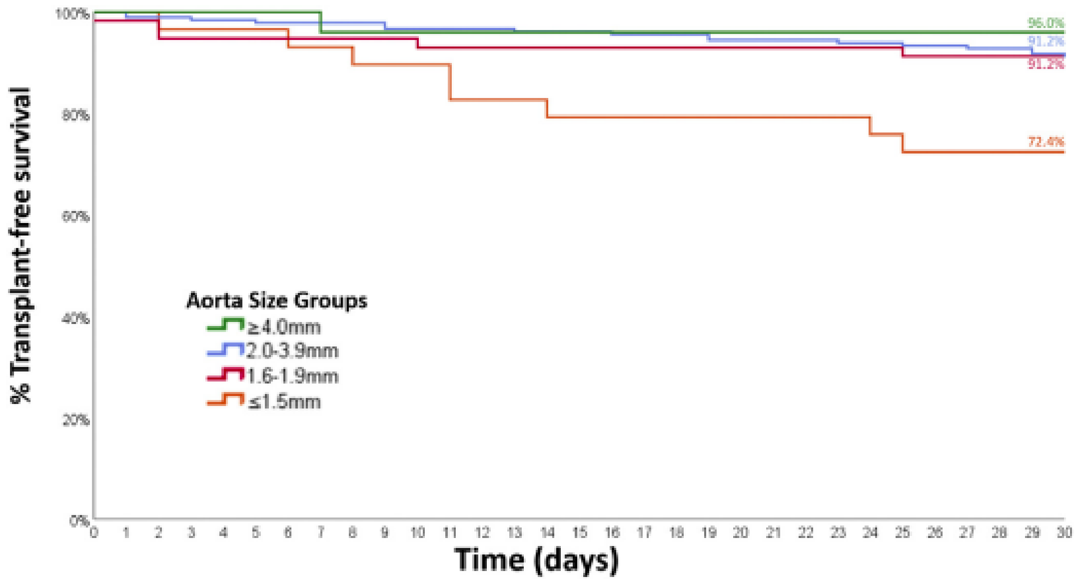

Figure 2. Transplant-free survival at 30 days post-Norwood by aortic diameter group (P = .008).

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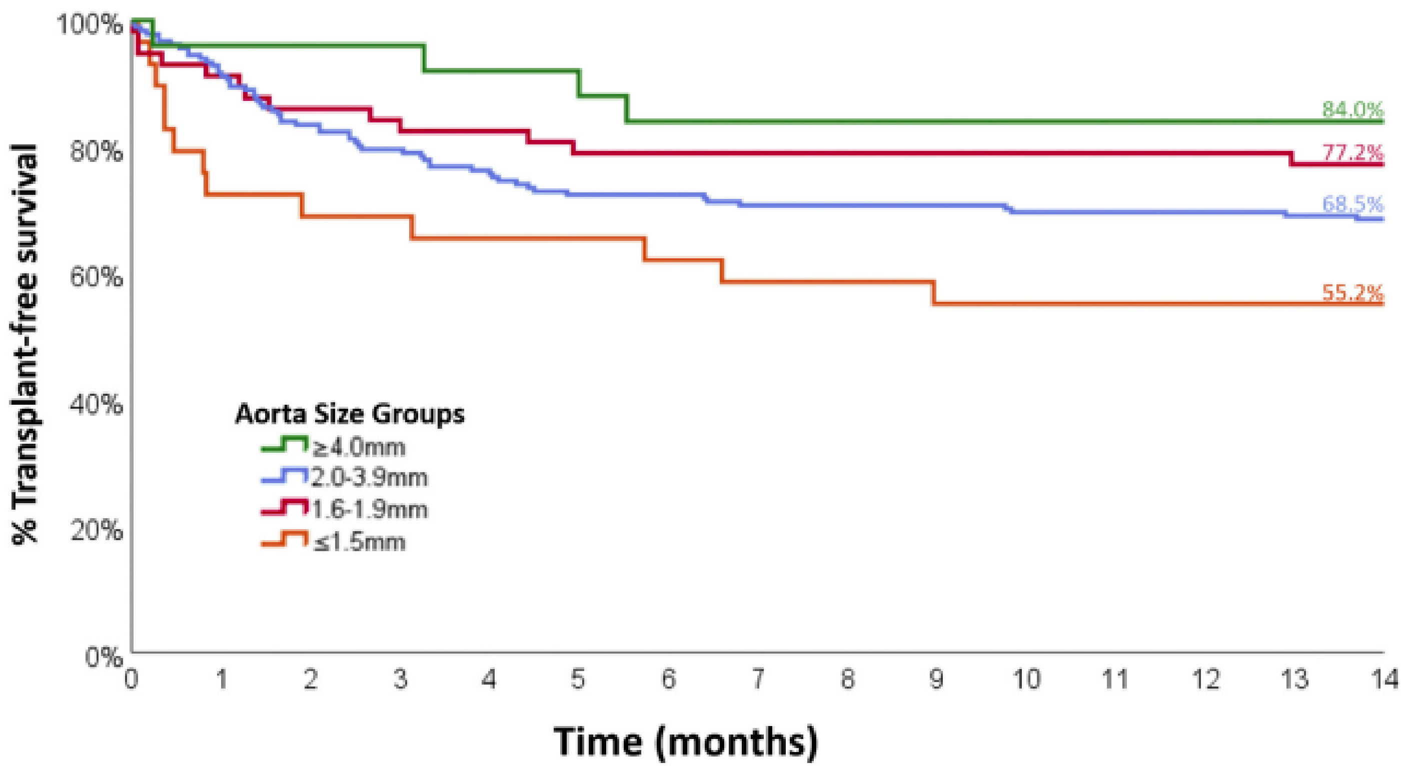

Figure 3. Transplant-free survival at 14 months post-Norwood by aortic diameter group (P = .052).

FACTORES DE RIESGO MORTALIDAD

- ✓ GENOPATIA
- ✓ PESO <2,5 kg
- ✓ PREMATUREZ

- ✓ SEPTUM INTACTO
- ✓ DRENAJE VENOSO ANOMALO PULMONAR TOTAL
- ❖ DISFUNCION DEL VD
- ❖ INSUFICIENCIA TRICUSPIDEA
- ❖ AORTA ASCENDENTE DIMINUTA