

**Online supplement - Table 1**

Children's cardiac diagnoses and previous surgical procedures, with classification at the time of the test, stating whether there was right to left shunt (actual or potential, including patients unrepaired/palliated), and their Hypoxic Challenge Test results.

<b>Gender</b>	<b>Age (yrs)</b>	<b>Diagnosis</b>	<b>Previous procedures</b>	<b>Classification</b>	<b>Baseline SpO<sub>2</sub></b>	<b>Test lowest SpO<sub>2</sub></b>
M	0.5	Coarctation of aorta; 1.1cm secundum ASD	Coarctation repair	Acyanotic with repair and potential for R-L shunt	96%	84% <i>FAIL</i>
M	1.8	Critical aortic stenosis; mitral stenosis	Aortic valve repair; mitral valve repair	Acyanotic with repair and no R-L shunt	100%	93%
M	3.3	Coarctation of aorta; bicuspid aortic valve; mitral stenosis	Coarctation repair; balloon dilatation of re-coarctation	Acyanotic with repair and no R-L shunt	100%	94%
F	0.8	Complete AVSD with large atrial & moderate ventricular component; moderate left AV regurgitation	Complete AVSD repair	Acyanotic with repair and no R-L shunt	100%	95%
M	2.1	Valvar & supra-valvar pulmonary stenosis; asymmetric septal LV hypertrophy; possible Noonan syndrome	Balloon dilation of pulmonary valve; surgical relief of pulmonary outflow stenosis (pulmonary valvotomy, infundibular myomectomy, patch enlargement of supra-valvar stenosis), enlargement of left PA, ASD closure	Acyanotic with repair and no R-L shunt	99%	94%
M	11.5	Anomalous origin of left coronary artery from anterior sinus with compression between aorta & pulmonary artery	Left internal mammary artery graft to left anterior descending coronary artery; enlargement of left coronary artery orifice; stenting of left coronary ostial stenosis	Acyanotic with repair and no R-L shunt	99%	91%
F	1.6	AVSD with large primum & ventricular components; Down syndrome	Complete AVSD repair	Acyanotic with repair and no R-L shunt	99%	89%
F	3.5	Pulmonary valvar & supra-valvar stenosis; mild right PA stenosis	Balloon dilation of pulmonary valve; surgical relief of RV outflow obstruction & patch enlargement of pulmonary trunk	Acyanotic with repair and no R-L shunt	99%	95%

M	5.5	Left atrial isomerism; biventricular AV connection; VA concordance; SVC & right azygous to right-sided LA; hepatic veins drain right sided LA; AVSD; small PDA; pulmonary stenosis	AVSD repair (re-op same day for patch dehiscence)	Acyanotic with repair and no R-L shunt	99%	92%
M	0.9	AVSD with separate valve orifices & isolated ventricular component (VSD); Down syndrome; prematurity with IUGR	Closure of VSD & closure of cleft of left AV valve. Operation note: no ASD at surgery	Acyanotic with repair and no R-L shunt	98%	88%
M	14.5	Sinus venosus defect	Repair sinus venosus defect	Acyanotic with repair and no R-L shunt	98%	95%
F	8.4	Coarctation of aorta; congenital mitral valve stenosis; bicuspid aortic valve with mild stenosis	Coarctation repair; mitral valve repair	Acyanotic with repair and no R-L shunt	98%	91%
M	6.3	Coarctation of aorta; partial AVSD; severe origin stenosis of right PA; Noonan syndrome. Tracheobronchomalacia with vocal palsy	Coarctation repair; balloon dilatation of right PA; closure of atrial communication & augmentation of right PA	Acyanotic with repair and no R-L shunt	97%	89%
F	2.8	Secundum ASD & PDA; tracheomalacia	Attempted device closure of ASD with device embolisation & urgent surgical retrieval of device with direct closure of ASD & PDA	Acyanotic with repair and no R-L shunt	97%	88%
F	0.5	Truncus arteriosus type I; ASD; moderate truncal valve regurgitation	Bilateral PA banding complete repair	Acyanotic with repair and potential for R-L shunt	98%	89%
M	1.5	Right coronary artery fistula to RV	None	Acyanotic without repair and no R-L shunt	100%	94%
M	1.6	Congenital aortic valve stenosis; moderate stenosis at present	None	Acyanotic without repair and potential R-L shunt	99%	92%
M	1.6	Tetralogy of Fallot	Tetralogy of Fallot repair - transannular patch with monocusp, closure of VSD, direct closure of PFO	Cyanotic with full repair and no R-L shunt	100%	94%
M	0.2	Transposition of great arteries	Balloon artery septostomy; arterial switch operation; ASD closure	Cyanotic with full repair and no R-L shunt	100%	92%

F	0.8	Total anomalous pulmonary venous connection to RA with restrictive PFO	Repair & reconstruction of atrial septum	Cyanotic with full repair and no R-L shunt	100%	94%
M	0.9	Critical pulmonary stenosis; currently mild (gradient 34mmHg)	Balloon dilation of pulmonary valve	Cyanotic with full repair and no R-L shunt	99%	94%
F	1.6	Transposition of great arteries with intact ventricular septum; moderate-severe proximal PA stenoses	Arterial switch operation; balloon PA angioplasty x2	Cyanotic with full repair and no R-L shunt	99%	93%
M	0.3	Transposition of great arteries	Arterial switch operation & ASD closure	Cyanotic with full repair and no R-L shunt	99%	89%
M	3.7	Tetralogy of Fallot with pulmonary atresia, accessory left anterior descending from right coronary	Left MBT shunt with left PA reconstruction; balloon angioplasty of left PA; complete repair of Tetralogy of Fallot; reconstruction of pulmonary bifurcation	Cyanotic with full repair and no R-L shunt	99%	92%
F	2.6	Transposition of great arteries with muscular outlet VSD; left PA stenosis	Arterial switch with VSD closure & PDA ligation; resection of supra-valvar aortic stenosis with left PA augmentation	Cyanotic with full repair and no R-L shunt	99%	93%
F	1.7	Transposition of great arteries; large ASD & aortopulmonary window	Arterial switch; closure of ASD & aortopulmonary window	Cyanotic with full repair and no R-L shunt	99%	94%
F	10.9	Tetralogy of Fallot with left anterior descending coronary artery anomaly (crossing RV outflow tract); right ventricular enlargement with pulmonary regurgitation & pulmonary stenosis	Tetralogy of Fallot repair with transannular patch & RV-PA conduit	Cyanotic with full repair and no R-L shunt	98%	91%
F	1.8	Tetralogy of Fallot & pulmonary atresia; PDA	Right MBT shunt; complete repair leaving PFO & 4mm residual VSD	Cyanotic with full repair and potential for R-L shunt	99%	76% <i>FAIL</i>
M	12.1	Hypoplastic left heart syndrome	Neonatal right MBT shunt; atrial septectomy; balloon dilatation of coarctation; bidirectional cavopulmonary anastomosis; fenestrated total cavopulmonary connection & augmentation of left PA; transcatheter closure of inter-atrial fenestration	Cyanotic with full repair and potential for R-L shunt	96%	90%
M	5.5	Situs inversus; left AV valvar atresia with indeterminate ventricle;	MBT shunt; bidirectional cavopulmonary anastomosis with left PA	Cyanotic with full repair and potential for	96%	83% <i>FAIL</i>

		presumed veno-venous collaterals (desaturates on exercise)	augmentation; completion of total cavopulmonary connection with extra-cardiac conduit; no fenestration	R-L shunt		
F	9.2	Absent right AV connection; concordant VA connections; PA stenosis	Central shunt & atrial septectomy; bidirectional cavopulmonary anastomosis with central shunt in situ; completion of total cavopulmonary connection	Cyanotic with full repair and potential for R-L shunt	91%	83%
F	1.5	Tetralogy of Fallot with severe pulmonary; branch PA stenoses	RV outflow tract resection with transannular patch & PDA ligation; permanent pacemaker; repair of Tetralogy of Fallot; ASD left open	Cyanotic with full repair and potential for R-L shunt	89%	74% <i>FAIL</i>
F	7.5	Mesocardia; situs solitus; absent right AV connection; single outlet heart with pulmonary atresia; RV connected to aorta; disconnected branch PAs	Neonatal right MBT shunt; left MBT shunt; bidirectional cavopulmonary anastomosis, reconstruction of PA, central shunt & ligation of previous shunts; fenestrated total cavopulmonary connection	Cyanotic with full repair and potential for R-L shunt	85%	78%
M	5.2	Severe Ebstein malformation of the tricuspid valve; moderate tricuspid regurgitation; ASD	Ebstein repair with Cone technique; small to moderate residual ASD	Cyanotic with full repair and potential for R-L shunt	80%	72%
F	0.5	Tetralogy of Fallot	Left MBT shunt	Cyanotic with palliated CHD	97%	90%
F	8.2	Situs inversus; AV & VA discordance; perimembranous inlet VSD; valvar & supravalvar pulmonary stenosis	Left MBT shunt; right MBT shunt; resection of subpulmonary stenosis; pulmonary valvotomy	Cyanotic with palliated CHD	97%	89%
F	1.7	Tetralogy of Fallot; non-confluent PAs; right PA arising from pulmonary trunk; left PA from arterial duct	Left MBT shunt & ligation of PDA; balloon angioplasty of left MBT shunt; surgical augmentation of RV outflow tract	Cyanotic with palliated CHD	96%	89%
M	1.9	Valvar pulmonary atresia with intact ventricular septum; dysplastic tricuspid valve with stenosis	Radiofrequency perforation & balloon dilation of pulmonary valve; right MBT shunt; revision of MBT shunt	Cyanotic with palliated CHD	94%	87%
F	1.9	AV discordance, double outlet RV with subpulmonary perimembranous outlet VSD; PDA	PDA ligation	Cyanotic with palliated CHD	92%	79%
M	1.5	Pulmonary atresia with VSD; MAPCAs; aortic	Unifocalisation of MAPCAs; RV-PA	Cyanotic with palliated CHD	92%	75% <i>FAIL</i>

		valve stenosis; 22q11 deletion	conduit; aortic valvotomy; augmentation of PA bifurcation			
F	9.3	Critical pulmonary stenosis; secundum ASD; long segment right PA hypoplasia/stenosis; severe pulmonary regurgitation	Balloon dilation of pulmonary valve; right MBT shunt	Cyanotic with palliated CHD	91%	85%
M	1.2	Double outlet RV with subaortic VSD; mid-muscular VSD	PA banding with PDA ligation	Cyanotic with palliated CHD	89%	79%
F	10.7	Double inlet indeterminate ventricle; pulmonary atresia; bilateral SVCs	Bilateral bidirectional cavopulmonary anastomosis	Cyanotic with palliated CHD	88%	82%
F	5.9	Double outlet RV; valvar & subvalvar pulmonary stenosis; non-committed VSD; ASD; MAPCAs	Right MBT shunt; bilateral bidirectional cavopulmonary anastomosis with right PA enlargement, ligation of pulmonary trunk & division of MBT shunt; angioplasty of right PA with atrial septectomy; epicardial pacemaker insertion	Cyanotic with palliated CHD	88%	82%
M	2.5	Tricuspid atresia with concordant VA connections	Right MBT shunt; bilateral bidirectional cavopulmonary connection with right PA reconstruction & reimplantation of shunt	Cyanotic with palliated CHD	87%	82%
M	0.8	Right atrial isomerism; unbalanced AVSD (dominant RV) with large ventricular & atrial components; double outlet RV; total anomalous pulmonary venous connection	Repair of total anomalous pulmonary venous connection & PA banding; tightening of PA band & bilateral bidirectional cavopulmonary anastomosis	Cyanotic with palliated CHD	87%	83%
M	2.0	Absent left AV connection; concordant VA connections; secundum ASD; perimembranous VSD; mid-muscular VSD; coarctation of aorta	Coarctation repair & PA banding; bidirectional cavopulmonary connection with aortopulmonary anastomosis (DKS), PA augmentation, atrial septectomy; transcatheter embolisation of aortopulmonary collateral	Cyanotic with palliated CHD	86%	76%
M	2.7	Situs solitus; double inlet RV; criss-cross AV connection; pulmonary atresia; ASD; left PA origin stenosis; RV dysfunction	Right MBT shunt & PDA ligation; left PA augmentation; bidirectional cavopulmonary anastomosis, BT shunt take down & atrial septectomy	Cyanotic with palliated CHD	84%	79%

F	4.2	Hypoplastic left heart syndrome	Norwood procedure; revision of RV to PA conduit; revision of RV to PA conduit & reconstruction of PA bifurcation; balloon angioplasty of both PAs; classical Glenn with disconnection PAs & RV to Sano left PA conduit	Cyanotic with palliated CHD	84%	75%
F	1.8	Pulmonary atresia with VSD & MAPCAs; confluent hypoplastic branch PAs; 22q11 deletion	Central shunt from left brachiocephalic artery to pulmonary trunk	Cyanotic with palliated CHD	83%	77%
F	2.7	Left atrial isomerism; biventricular AV connection; interrupted IVC with azygous continuation to right SVC; hepatic veins drain to right sided LA; pulmonary veins to left sided LA	Left PA balloon dilation	Cyanotic with palliated CHD	83%	73%
F	1.0	Double inlet LV, discordant VA connection; right sided rudimentary RV; non-restrictive VSD; pulmonary stenosis	Balloon atrial septostomy; bidirectional cavopulmonary anastomosis, atrial septectomy & closure of pulmonary trunk	Cyanotic with palliated CHD	83%	71%
F	2.7	Tricuspid atresia with concordant VA connections; pulmonary stenosis	Right MBT shunt; bidirectional cavopulmonary anastomosis, take down of MBT shunt, atrial septectomy	Cyanotic with palliated CHD	82%	73%
M	1.4	Transposition of great arteries with subvalvar & valvar pulmonary stenosis & subpulmonary VSD	Left MBT shunt & PDA ligation; urgent right MBT shunt	Cyanotic with palliated CHD	82%	76%
M	1.2	Pulmonary atresia; VSD	Left MBT shunt; right MBT shunt	Cyanotic with palliated CHD	82%	75%
M	2.5	Tetralogy of Fallot with pulmonary atresia; MAPCAs; hypoplastic confluent PAs	Right MBT shunt; RV-PA conduit & occlusion MBT shunt; right PA device occlusion of right MAPCA	Cyanotic with palliated CHD	82%	75%
M	1.5	Tetralogy of Fallot with pulmonary atresia; possible Mowat Wilson syndrome	Neonatal right MBT shunt; left MBT shunt	Cyanotic with palliated CHD	80%	74%
F	0.9	Pulmonary atresia with VSD & MAPCAs; hypoplasia of left PA	Left MBT shunt, unifocalisation of lung blood supply	Cyanotic with palliated CHD	80%	72%

M	0.9	Pulmonary atresia with intact ventricular septum; coronary fistula	Right MBT shunt; bidirectional cavopulmonary anastomosis & atrial septectomy & closure of MBT shunt	Cyanotic with palliated CHD	79%	74%
M	1.2	Tricuspid atresia with concordant VA connections	Atrial septectomy & bidirectional cavopulmonary anastomosis	Cyanotic with palliated CHD	78%	69% <i>FAIL</i>
F	3.0	Transposition of great arteries with VSD & pulmonary stenosis	Right MBT shunt; left MBT shunt	Cyanotic with palliated CHD	76%	73%
M	0.3	Ebstein malformation of the tricuspid valve; ASD	None	Cyanotic with unrepaired CHD	96%	88%
M	0.4	Severe Ebstein malformation of the tricuspid valve	None	Cyanotic with unrepaired CHD	96%	90%
M	3.1	Ebstein malformation of the tricuspid valve	None	Cyanotic with unrepaired CHD	96%	85% <i>FAIL</i>
M	0.7	Complete AVSD with Tetralogy of Fallot; Down syndrome	None	Cyanotic with unrepaired CHD	88%	81%
F	6.4	Ebstein malformation of tricuspid valve	None	Cyanotic with unrepaired CHD	86%	77%
F	4.0	Ebstein malformation of tricuspid valve; large secundum ASD; right aortic arch with aberrant left subclavian artery (vascular ring substrate)	None	Cyanotic with unrepaired CHD	82%	75%
F	0.9	Left atrial isomerism; hepatic vein to left sided LA; concordant VA connections; dextrocardia; pulmonary veins to left sided LA, interrupted IVS with azygous continuation to left SVC to left sided LA; common atrium with AVSD; no ventricular component; moderate left AV regurgitation; pulmonary valvar stenosis; PDA	None	Cyanotic with unrepaired CHD	81%	66% <i>FAIL</i>

**Abbreviations:** ASD: atrial septal defect; AV: atrioventricular; AVSD: atrioventricular septal defect; CHD: congenital heart disease; DKS: Damus-Kaye-Stansel procedure; HCR: hypoxic challenge test; IUGR: intrauterine growth restriction; IVC: inferior vena cava; LA: left atrium; LV: left ventricle; MAPCAs: multiple aortopulmonary collaterals; MBT: modified Blalock-Taussig; PA: pulmonary artery; PDA: patent ductus arteriosus; PFO: patent foramen ovale; R-L: right to left; RA: right atrium; RV: right ventricle; RV-PA: right ventricle to pulmonary artery; SpO<sub>2</sub>: arterial oxygen saturation measured by pulse oximetry; SVC: superior vena cava; VA: ventriculo-arterial; VSD: ventricular septal defect.

**Online supplement - Table 2**

Baseline data and Hypoxic Challenge Testing results for those who passed the test (n=60) vs those who failed the test (n=8). Results are expressed as medians (IQR) unless stated otherwise. Data refer to the total patient numbers for the two groups except where stated.

<b>Median (IQR)</b>	<b>Passed test</b>	<b>Failed test</b>	
No.	60	8	
Male n (%)	32 (53%)	4 (50%)	
Age (yrs)	2.0 (1.2-4.5)	1.3 (0.9-1.6)	ns
SpHb (g/dl)	12.3 (10.8-13.0) n = 32	12.7 (11.9-13.9) n = 8	ns
Baseline SpO <sub>2</sub> (%)	96 (86-99)	91 (83-96)	ns
Lowest test SpO <sub>2</sub> (%)	88 (78-92)	75 (69-78)	p=0.001
Absolute drop in SpO <sub>2</sub> (%)	7 (6-8)	15 (13-16)	p<0.0001
Time to reach lowest SpO <sub>2</sub> (mins)	7.0 (5.0-10.0) n = 34	5.4 (5.1-5.9) n = 7	ns
Baseline heart rate (bpm)	118 (97-134)	133 (122-137)	ns
Test heart rate (bpm)	118 (99-133)	133 (127-139)	ns
Change in heart rate (bpm)	1 (-6 to +7)	5 (+3 to +10)	ns
Baseline PtcCO <sub>2</sub> (kPa)	4.3 (4.0-4.5) n = 58	4.2 (3.7-4.8) n = 8	ns
Test PtcCO <sub>2</sub> (kPa)	4.3 (3.9-4.6) n = 57	4.4 (3.9-4.9) n = 8	ns
Change in PtcCO <sub>2</sub> (kPa)	0 (-0.2 to +0.2)	0 (-0.4 to +0.2)	ns
Baseline QTc (ms)	410 (394-426) n = 53	406 (397-431) n = 6	ns
Test QTc (ms)	419 (404-426) n = 53	422 (396-429) n = 6	ns
Change in QTc (ms)	+2 (-4 to +21) n = 53	0 (-3 to +16) n = 6	ns