Adult CHD – Success Leads to Problems Prof Kevin P Walsh, MD FRCPI

UCD Clinical Professor

Consultant Cardiologist, Congenital Heart Disease



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THIS IS MODERN MEDICINE

It starts like this





It might end like this



Because of this





Due to:





The Difficulty: RVOT expansion and No Landing Zone



Represents 85% of dysfunctional RVOTs

Consequences of Post-op Fallot Pulmonary Regurgitation

- Right Ventricular Dilation
- Effort Intolerance/CCF
- Arrhythmias/Sudden death
- Multiple pulmonary valve replacements



Pacemaker ICD Therapy



RF Energy On



Bonhoeffer Valve

(Melody Valve and Ensemble Delivery System)





Re-Valving



Venus P-Valve



RVOT



Venus Valve-Dublin Case



Delivery to LPA



Pullback



Full Deployment



Post







TETRALOGY OF FALLOT Long term outcome



Murphy JG et al. NEJM 1993;329(9): 593 Bertranou EG. et al. Am J Cardiol 1978;42:458.

The problem of complex defects....





The problem of complex defects....





Separation of the Circulations



Role of Right (Sub-Pulmonary) Ventricle



The Fontan Pressurizes the Liver





Types of Fontan





Freedom from death or transplantation according to type of Fontan.



Paul Khairy et al. Circulation. 2008;117:85-92

A Fontan DOB 1976

- Atrial tachycardias were the main issue pre-conversion with amiodarone thyrotoxicosis requiring thyroidectomy in 1996
- Had A Fib post Fontan conversion and was on Amiodarone 400 mg OD thereafter
- Developed corneal opacification in Leye requiring corneal transplant under GA
- CT-Liver in 2014 showed a calcified 2.8 cm cyst, nodular shrunken liver and gall stones
- Echo in 2015 showed increased OFO (Sub Aortic stenosis) @ 4.5m/s

A Fontan DOB 1976

Cath 2015

- IVC mean 8
- LPA mean 7 (11 after dye)
- RPA mean 7
- Wedge mean 3
- LV 164/8 gradient 60 mmHg





Pediatric Congenital Heart Disease (G Singh, Section Editor)

Fontan Liver Disease: Review of an Emerging Epidemic and Management Options

Elisa Bradley, MD^{1,2,*} Benjamin Hendrickson, MD^{1,2} Curt Daniels, MD^{1,2}

Combined Heart and Liver Transplantation



A Fontan 1976

- Transplant MDT listed for OHT in March 2016
- Waited
- OHT on 1/1/18 (LN)
- Returned to work 9/2018

60% Increase in Adult Prevalence of Single Ventricles by 2023



- prevalence 1960's







Older adult



Young adult

Congenital Heart Disease - prevalence 1990's







Older adult

Child

Young adult

Congenital Heart Disease - prevalence 2023







child

Young adult

Older adult

Congenital Heart Disease - prevalence 2028











Older adult

BETHESDA MODEL

32nd Bethesda Conference: Care of the adult with congenital heart disease JACC 2001;37(5)1161-1198

Birth rates 1940 - 1989 complex CHD 1.5 per 1,000 live births simple 4.5 per 1,000 live births

 Estimated likely survival rates for birth epoch (from literature)

Simple

- Secundum atrial septal defects
- Small ventricular septal defects
- Mild pulmonary stenosis
- Bicuspid aortic valve/Mild AS-AI
- Surgically Closed VSD

Complex

- Single Ventricles
- Truncus
- Atrioventricular septal defects
- Transposition
- Double outlets
- Fallot's tetralogy
- Pulmonary atresia
- Ebstein's anomaly
- Anomalous pulmonary veins
- Cyanotics

Survival of Complex CHD

32nd Bethesda Conference: Care of the adult with congenital heart disease JACC 2001;37(5)1161-1198

Era	1 st Year Survival Rate	18 yrs Survival Rate	
1940-1960	20%	10%	
1960-1980	50%	35%	
1980-1990	70%	50%	
1990-2000	85%	70%	

Adult CHD in Eire - Numbers

Era	Births per	Incidence		Survival to 18 years	
	year	Complex	Simple	Complex	Simple
1940- 1960	63,565	95/y = 1900	285/y = 5700	190 (10%)	5150 (90%)
1960- 1980	60,735	91/y = 1820	273/y = 5460	640 (35%)	4920 (90%)
1980- 1990	74,064	111/y = 1110	333/y = 3330	555 (50%)	3000 (90%)
1990- 2000	53,044	80/y = 800	240/y = 2400	680 (70%)	2160 (90%)
		Prevalence in 2020		2,065	15,210
		2023		2,421	16,411

Conclusions

- The successful treatment and survival of patients with congenital heart disease has created a new group of patients
- They require specialised follow-up and often complex multidisciplinary management and interventions

ADULTS WITH CONGENITAL HEART DISEASE



"Science is always wrong. It never solves a problem without creating ten more.....

George Bernard Shaw

Thank you

