Conduction Disturbance in Congenital Heart Disease

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Overview

- Conduction system in CHD
- Normal conduction system
- Congenital heart disease

VSD	ASD
AVSD	TAPVC
Ebstain Anomaly	
TOF	Floppy Mitral Valve/MVP
TGA	Corrected TGA
HLHS	DORV
DILV	Isomerism

- Arrhythmias after CPB

Introduction

Conduction system in congenital heart disease

Organogenesis defect

Congenital defect ->

conduction system : migration, interruption

Volume overload – chamber enlargement

conduction delay, interruption

Pressure overload- hypertrophy, fibrosis

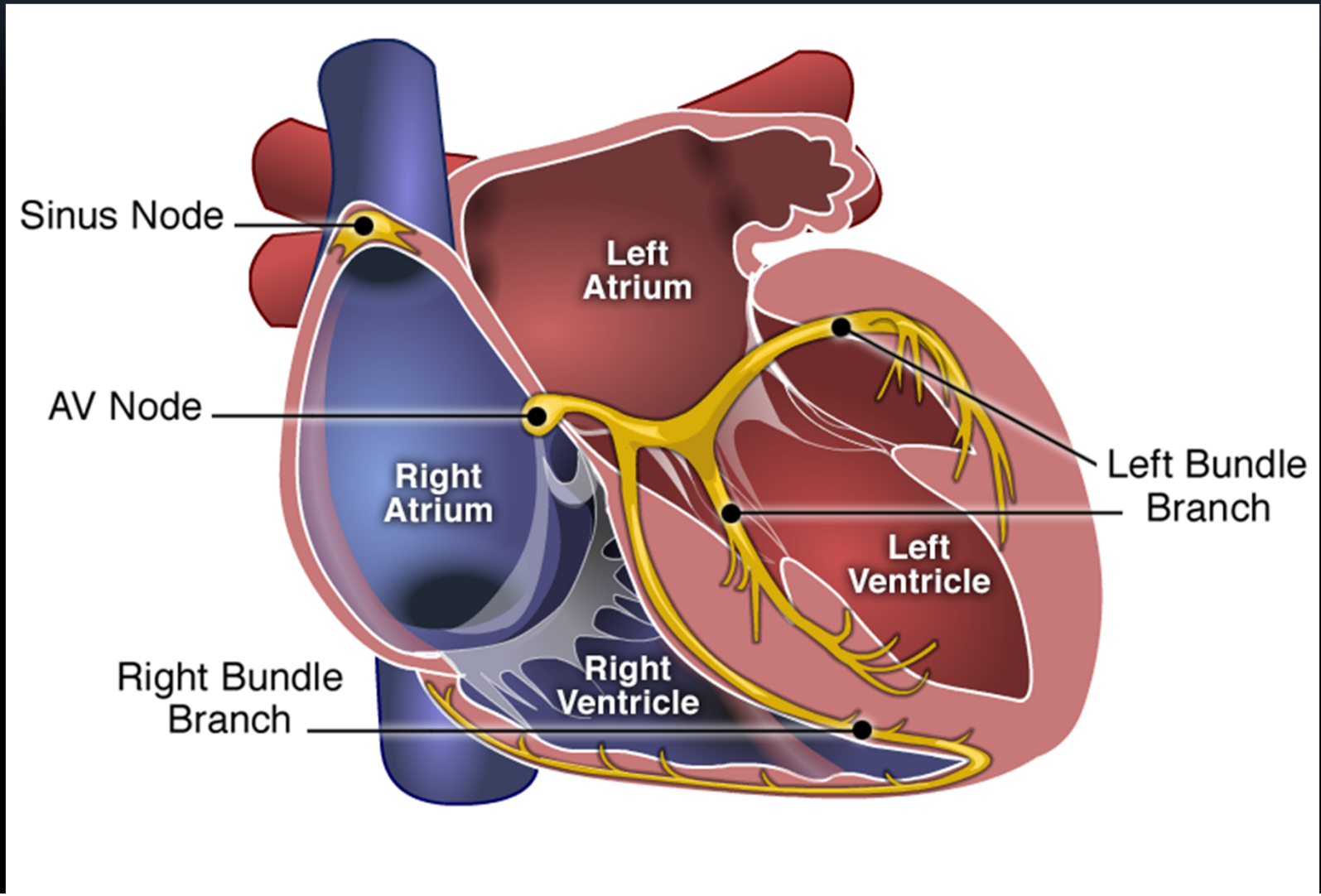
Operation associated conduction abnormality

conduction ts direct injury

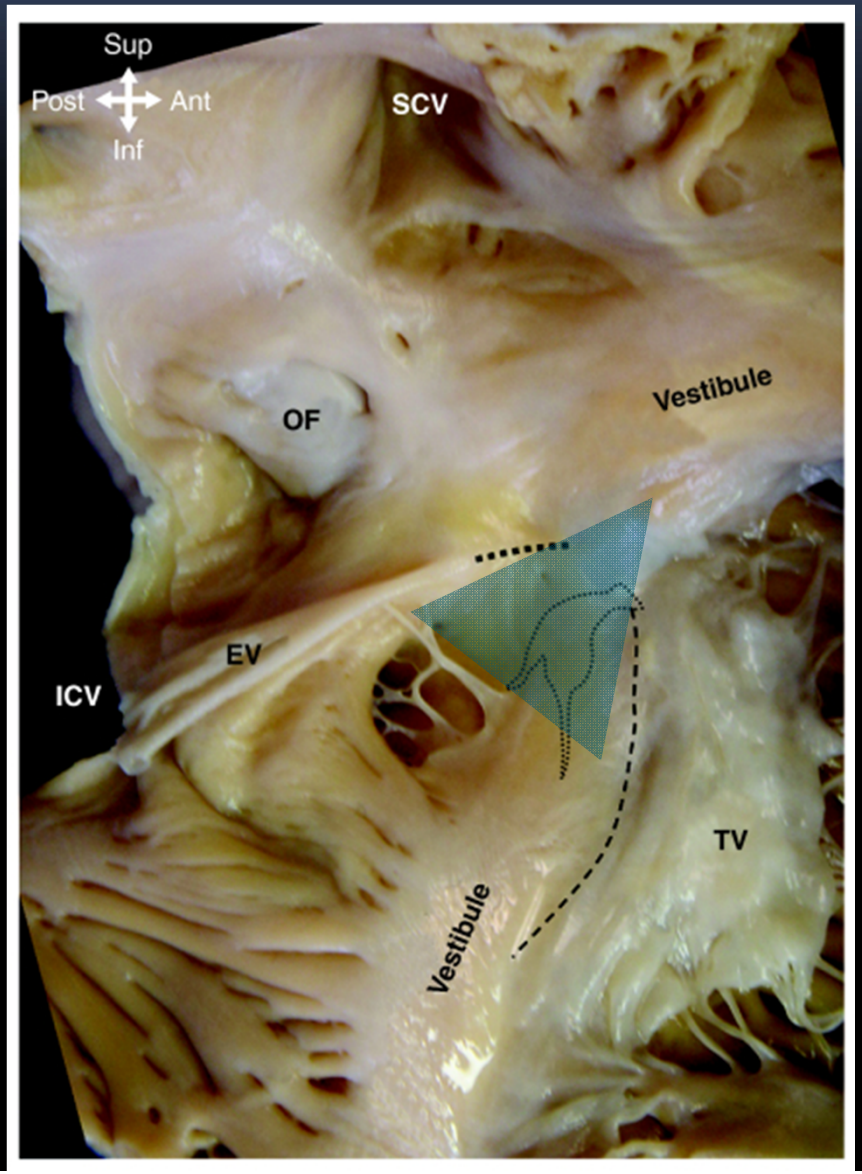
edema

hemorrhage

Normal conduction

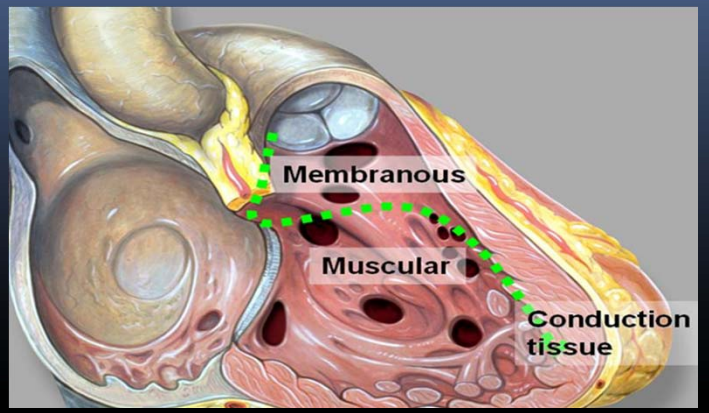


AV node



Triangle of Koch
- base of the septal leaflet of TV,
- AM margin of CS orifice,
- palpable subendocardial collagen bundle (tendon of Todaro)

Ventricular Septal Defect



AV conduction to Defect -relationship

important to surgical repair

His bundle

PM : along the posterior-inferior margin of the defect.

Inlet : antero-superiorly to the defect

Anderson et al. J Cardiac Surg 1992;7:17–34.

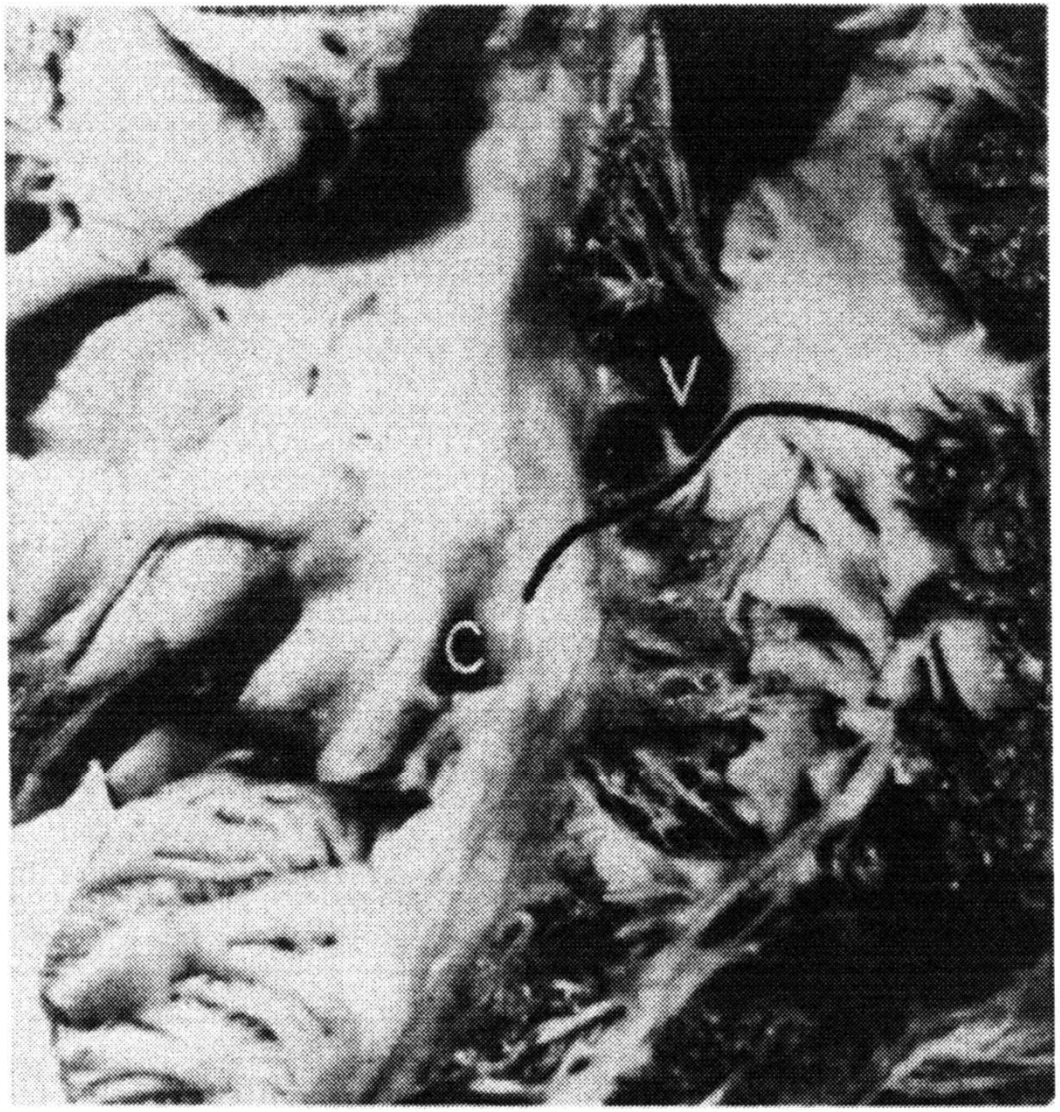
Muscular & outlet, SA : little danger of heart block

extend into the PM : may occur

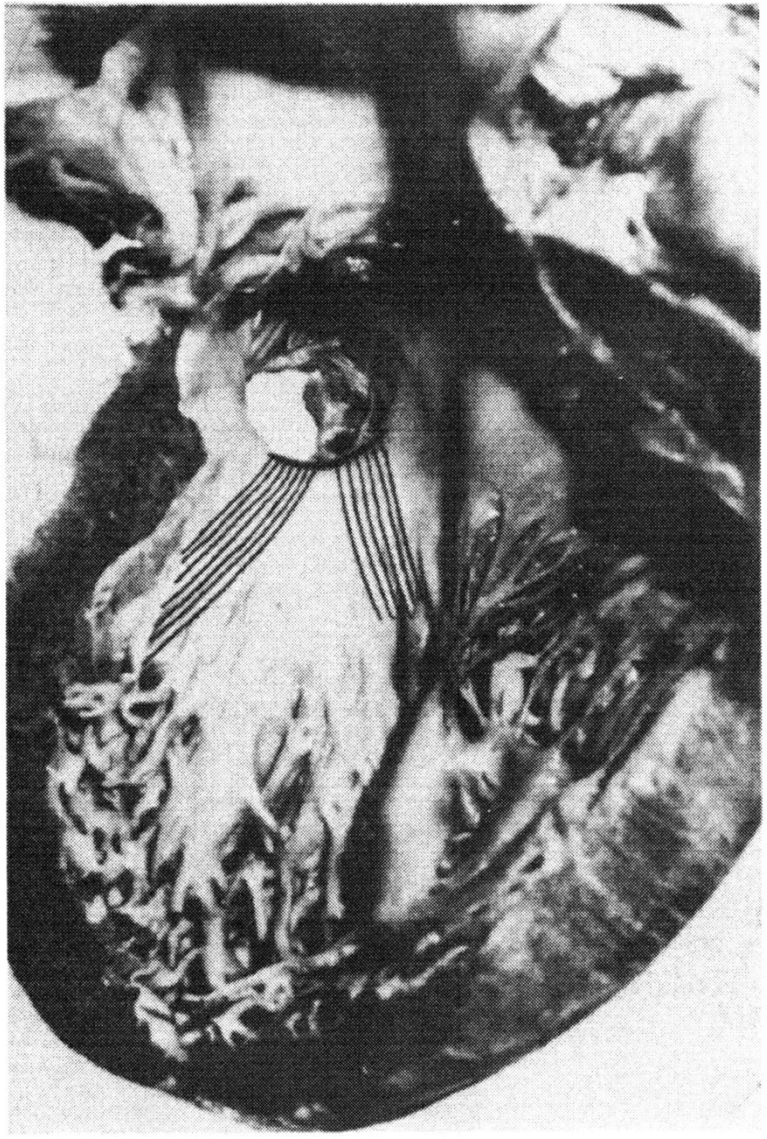
RBBB after repair

ventriculotomy or direct injury of Rt bundle

Allen et al. Moss and Adams' heart disease in infants, children, and adolescents. 2008.



A



B

Lev M. Am J Cardiol. 1968 May;21(5):619-27.

Atrial Septal Defect

PR interval : prolonged in older patients

intra-atrial & sometimes H-V conduction delay

→ 1st AV block

Shiku DJ et al. J Electrocardiol 1982;15:9-14

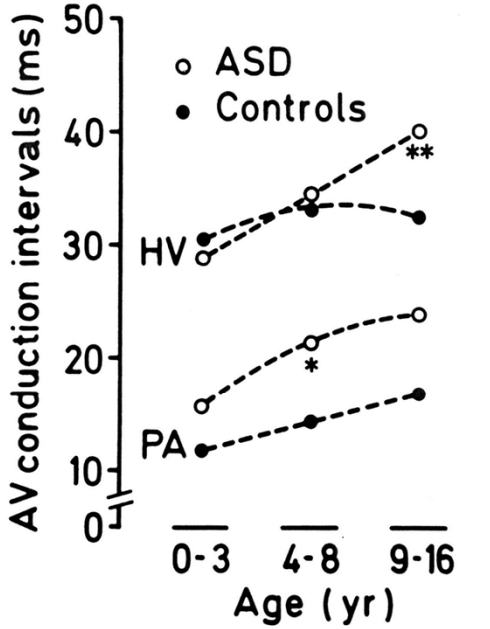


Fig. 2. Changes of PA and HV intervals with age in children with and without ASD. Open and filled circles represent mean values of ASD and control subgroups respectively. * ASD value is significantly different from the corresponding control value at $P < 0.05$, ** at $P < 0.01$.

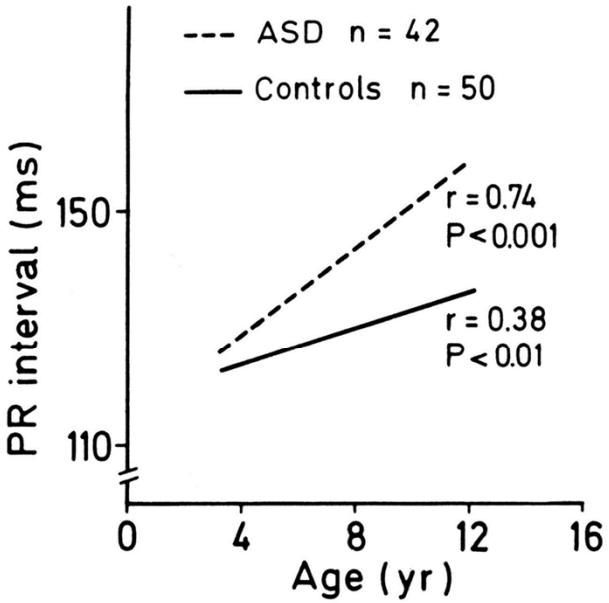


Fig. 3. Linear regression of the PR interval in relation to age in children with and without ASD. The slope of the PR line in ASD patients is significantly steeper than that of the PR line in the control group ($P < 0.05$).

Sinus node dysfunction by EPS

corrected sinus node recovery times

sinoatrial conduction times → abnormal

rare in resting ECGs or 24-hour ambulatory monitoring

→ predisposed to atrial arrhythmias

Clark EB et al. Circulation 1982;65:976-980

Karpawich Pp et al. Am J Cardiol 1985;55:519-521

Bink-Boelkens MTE et al. Int J Cardiol 1988;20:263-272

TABLE 1. *Electrophysiologic Data of Preoperative Patients with Secundum Atrial Septal Defects*

Pt	Age (years)	Sex	Symptoms	ECG ARR	24-hour ECG ARR	Intracardiac electrophysiology: Sinoatrial node CSNRT (msec)	Sinoatrial node SACT (msec)	AV node AH (msec)	AV node HV (msec)	AV node W@APR (beats/min)
1	13	F	—	1° AV block	Sinus arrest junctional	~40	—	180	40	153
2	13	F	Syncope		2° AV block SAN arrest	800	—	80	45	140
3	18	M	—	—	Normal	450	286	95	53	120
4	4 6/12	F	—	—	Normal	470	158	60	35	—
5	6 6/12	M	—	—	NA	110	130	70	40	200
6	3 9/12	M	—	—	NA	80	126	70	35	200
7	6 3/12	M	—	—	Paroxysmal atrial flutter	SAN-EB	SAN-EB	—	—	200*
8	1 4/12	F	—	—	NA	130	—	80	40	200
9	13	F	—	—	2 episodes Mobitz II AV block	280	140	75	50	140
10	10	M	Dizziness	—	Normal	340	174	90	45	150
11	10 6/12	M	—	—	Wandering atrial pacemaker	390	—	80	40	200
12	1 6/12	F	—	—	NA	130	122	85	30	200
13	10/12	F	—	—	Frequent PACs	330	168	90	35	200
14	16	M	—	—	Normal	440	300 intermittent SAN-EB	90	40	200
15	1 4/12	F	—	—	Normal	120	140	60	35	200
						Normal[6] < 275	Normal[6] < 200	Normal[8] < 120	Normal[8] < 55	Normal[9] > 160

*Wenkebach assumed to be in AV node because His could not be recorded.

Abbreviations: @CL = obtained at sinus cycle length; AH = atrial-His duration; ARR = arrhythmias; AV = atrioventricular; CSNRT = corrected sinus node recovery time; ERP = effective refractory period; FRP = functional refractory period; % green dye = left→right shunt calculated from green dye curves; HV = His to ventricular duration; NA = not available; Qp/Qs = pulmonary-to-systemic flow ratio; S = surgical repair; SACT = sinoatrial conduction time; SAN = sinoatrial node; SAN-EB = SAN entrance block; W@APR = atrial paced AV node Wenckebach rate; PACs = premature atrial complexes.

Clark EB et al. Circulation 1982;65:976-980

ECG : 40 ASD

6 (15%) 1st AV block -> 5/6 disappear after Op.

TABLE 2

Pre- and postoperative 24-hour Holter recordings.

	Preoperative $n = 31$	Postoperative $n = 31$
Abnormal Holter recordings	14	8
Accelerated atrial rhythm	11	6
AVJ rhythm	1	2
Sinus arrest	2	–
2nd degree AV block		
type I	1	–
type II	2	–
PACs > 10/hr	1	1
PVCs > 10/hr	–	1

PAC = premature atrial contraction; PVC = premature ventricular contraction; AVJ = atrioventricular junctional.

Bink-Boelkens MTE et al. Int J Cardiol 1988;20:263-272

TABLE I Electrophysiologic Data

	Preoperative Abnormal Electrophysiology							Postoperative						
					Atrium							Atrium		
Age (yr)	CL	CSNRT	SACT	WCL	FRP	ERP	AVN FRP	CL	CSNRT	SACT	WCL	FRP	ERP	AVN FRP
2	680	300	EB	400	260	240	320†	600	360	122	280	240	230	290†
3	650	350	180	360										
4	630	360	EB	320	310	300	340							
4	610	400	112	320	290	260	270	550	300	204	260	240	230	270†
4	700	450	112	500	270	240	310	680	390	268	400	240	220	250
4	650	380	206	280	310	300	320	590	330	136	280	220	170	260†
4	650	390	198	340	240	230	250							
5	610	320	160	350	250	240	260							
10	970	450	332	550	310	300	440	860	370	138	500	290	280	410
16	810	450	134	320	290	280	300	710	310	184	300	300	280	310
17	850	340	182	520	320	300	310	700	490	204	400	230	200	280
Normal		<275	<210	<380	*	*	*							
	Preoperative Normal Electrophysiology													
1	510	180	80			270								
1	580	160	150											
2	600	230	80	350		210								
2	610	250	122	320	230	220	300							
3	500	60	164	360	190	180	310							
3	600	260	160	300	260	240	280							
3	520	260	192	280	260	240	290							
4	670	90	126	250	180	160	210							
5	640	220	168	380	200	190	260							
16	550	270	78	280	230	180	260							

All values are in milliseconds.
* Cycle length-dependent.
† Inducible supraventricular tachycardia.
CL = cycle length: CSNRT = corrected sinus node recovery time; EB = entrance block; ERP = effective refractory period; FRP = functional refractory period; SACT = sinoatrial conduction time; WCL = Wenckebach CL.

Karpawich Pp et al. Am J Cardiol 1985;55:519-521

Atrioventricular Septal Defect

AV node

displaced posteriorly, near the orifice of the coronary sinus

His bundle

displaced inferiorly, along the inferior rim of the septal defect

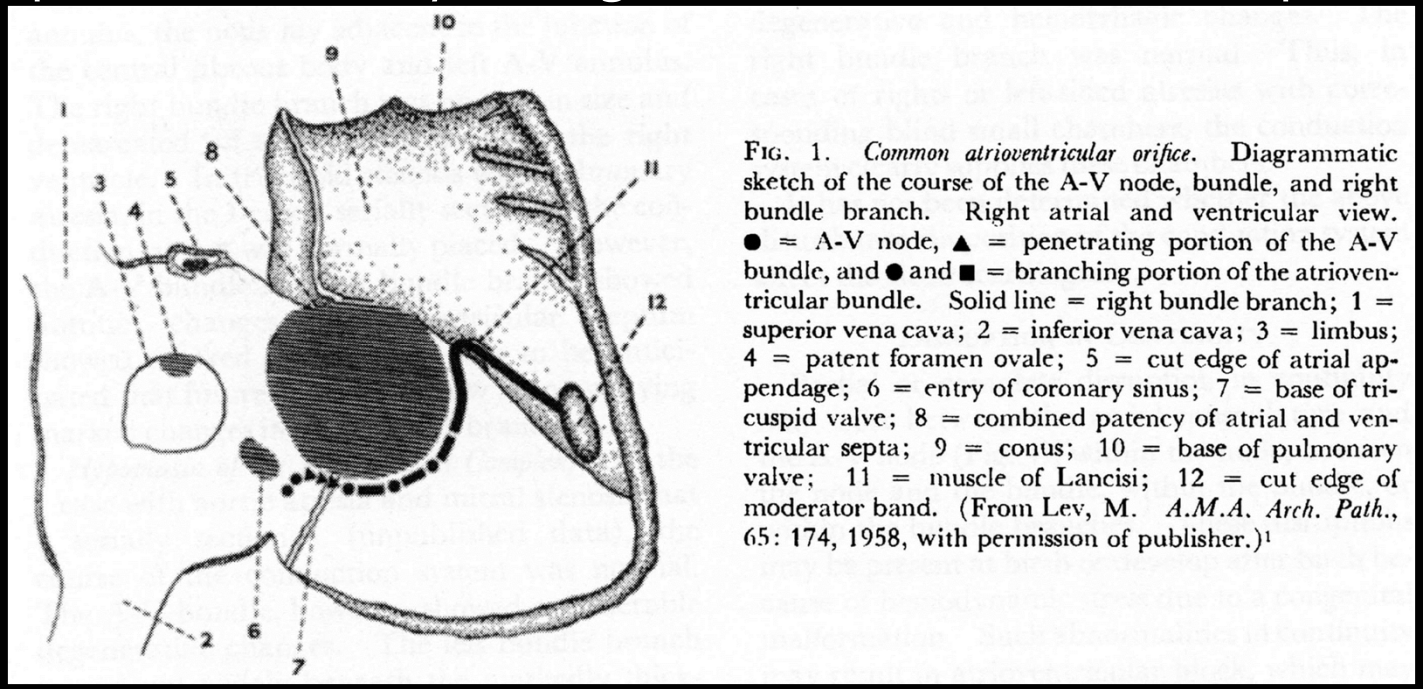


FIG. 1. *Common atrioventricular orifice.* Diagrammatic sketch of the course of the A-V node, bundle, and right bundle branch. Right atrial and ventricular view. ● = A-V node, ▲ = penetrating portion of the A-V bundle, and ● and ■ = branching portion of the atrioventricular bundle. Solid line = right bundle branch; 1 = superior vena cava; 2 = inferior vena cava; 3 = limbus; 4 = patent foramen ovale; 5 = cut edge of atrial appendage; 6 = entry of coronary sinus; 7 = base of tricuspid valve; 8 = combined patency of atrial and ventricular septa; 9 = conus; 10 = base of pulmonary valve; 11 = muscle of Lancisi; 12 = cut edge of moderator band. (From Lev, M. *A.M.A. Arch. Path.*, 65: 174, 1958, with permission of publisher.)[1]

Lev M. Am J Cardiol. 1968 May;21(5):619-27.

P-R interval prolongation

patient's age & HR (18 - 70%)

d/t intra-atrial conduction time

high right atrium ~ low septal right atrium

other intracardiac EPS measurements : normal

TABLE I Electrocardiographic Findings in Patients with Atrioventricular Canal

	Preoperative			Postoperative		
	CAVC	IAVC	Total	CAVC	IAVC	Total
PR	8/15	3/14	11/29	8/14	4/10	12/24
QRS (RBBB)	1/15	5/14	6/29	11/15	8/10	19/25
LAD	13/15	10/14	23/29	13/15	8/10	21/25
Rhythm Abn	0/15	0/14	0/29	2/16	0/10	2/26

Abn = abnormality; CAVC = complete atrioventricular canal; IAVC = incomplete atrioventricular canal; LAD = left-axis deviation; RBBB = right bundle branch block.

Fournier A, Young M-L et al. Am J Cardiol 1986;57:1137-1141

TABLE II Electrophysiologic Findings in Preoperative Patients

Pt	Associated Anomalies	Age (yr)	Basic Intervals						Sinus Node		Atrium		AV Node			Ventricle	
			PA	AH	HV	AV	PR	V-RV	SNRT	SACT	ERP	FRP	ERP	FRP	W	ERP	FRP
							Complete AV Canal										
1	PDA, ASD 2	1 2/12	30	34	36		100	26	360		140	180	ATL	ATL		170	185
2		6/12	10			124	134	0	550		175	180	ATL	ATL	<300	180	190
3	ASD 2	2 11/12	14	105*	32		151*	22		73	160	210	ATL	ATL			
4		2 4/12	20	70	44		134	14	420		160	205	ATL	ATL		180	190
5		9 2/12	20	150*	30		200*	0	900	120	180	265	ATL	ATL	300	230	240
6	PDA	9 4/12	44	101	31		176*	11	1,005	168	160	245	270	<350		210	220
7		3 2/12	47*	65	31		143†	25	610		210	230	250	340	310	250	260
							Incomplete AV Canal										
8		16 3/12	69*	76	42		187*	18	740		150	290	ATL	ATL	285	240	250
9		2 6/12	25	70	40		135	8	550	91	160	225	ATL	ATL	310		
10		12 3/12	39	66	41		146	53*	830	194	180	190	ATL	ATL	<300	190	210
11		3 6/12	51*	61	16		128†	20	660		175	210	210	210	<310	220	225
12		2 4/12	31	73	25		129	15			140	170	170	200			
13		6 1/12	37	67	37		141	15	1,230*	221*	200	220	ATL	ATL	<270	250	270
14		9/12	51*	76	16		143†	28	535		125	180	225	260	<280		
15		1 9/12	35	91	40		166*	20	410		135	165	ATL	ATL		230	250
16		3	23	83	37		143	10	608	90	180	240	ATL	ATL	350	200	220
17		3 9/12	24	75	29		128	30			<240	220	F270 S <240	F300		200	230
18	VSD, ASD 2	8/12	40*	50	40		130†	10	510		170	200	ATL	ATL	225	180	190

All intervals are in milliseconds.

* Abnormal value, † Normal PR but prolonged PA.

AS = aortic stenosis; ASD 2 = atrial septal defect secundum; ATL = atrial limited; AV = atrioventricular; ERP = effective refractory period; F = fast AV nodal pathway; FRP = functional refractory period; PDA = patent ductus arteriosus; PS = pulmonary stenosis; R Arch = right aortic arch; S = slow AV nodal pathway; SACT = sino atrial conduction time; SNRT = sinus node recovery time; V-RV = right ventricular apical activation time; VSD = ventricular septal defect type AV canal; W = Wenckebach periodicity.

Fournier A, Young M-L et al. Am J Cardiol 1986;57:1137-1141

Total Anomalous Pulmonary Venous Connection

RV volume overload

occasionally incomplete RBBB

Ebstain Anomaly

RAE – peaked P

P wave : more wider & notched (poor Px)

Wolff–Parkinson–White syndrome

20- 30%

m.c. : right-sided accessory pathway – LAD of delta wave

PSVT can be occurred

W/o Wolff–Parkinson–White syndrome

QRS complexes : RBBB

QRS interval : ↑ d/t intraventricular conduction delay

Arrhythmia

PSVT : w/wo WPW

atrial flutter or atrial fibrillation : older, severe

Other : wandering pacemaker or ventricular ectopy

Kumar AE, et al. Am J Cardiol 1971;28:84–95.

55 Ebstein anomaly

TABLE IV

Arrhythmias Observed in 24 Patients

Type of Arrhythmia	no. of Cases
Paroxysmal atrial tachycardia	7
Paroxysmal atrial tachycardia and WPW "B"	5
WPW "B"	3
Atrial flutter	2
Atrial fibrillation	1
Nodal rhythm	3
Wandering pacemaker	1
Premature ventricular contraction	2

WPW "B" = Wolff-Parkinson-White syndrome, type B.

TABLE V

Follow-up of the Electrocardiogram in 26 Patients with Ebstein's Anomaly with Time Interval of 5 or More Years

	P Amplitude (mm) (mean ± SEM)	P Duration (sec) (mean ± SEM)	P–R (sec) (mean ± SEM)	P/PQ Index (mean ± SEM)	QRS Duration (sec) (mean ± SEM)
First visit	2.8 ± 0.24	0.07 ± 0.004	0.15 ± 0.005	0.92 ± 0.05	0.09 ± 0.005
Last visit	3.8 ± 0.27	0.09 ± 0.004	0.19 ± 0.008	0.88 ± 0.04	0.12 ± 0.005
Mean difference	1.0 ± 0.16	0.02 ± 0.005	0.04 ± 0.009	−0.04 ± 0.06	0.03 ± 0.006
P value	<0.001	<0.01	<0.001	NS	<0.001

NS = not significant.

Kumar AE, et al. Am J Cardiol 1971;28:84–95.

Tetralogy of Fallot

In postoperative
- m.c. RBBB, early and late arrhythmias

In older children & adult untreated TOF
- ventricular ectopy & other arrhythmias
- d/t RV hypertension and myocardial fibrosis.

Sudden death & ventricular arrhythmias
- early in the experience with total repair of TOF
- risk : 4.6%
- d/t bi-fascicular block -> complete AV block
  - re-emergence of transient postOp complete AV block
  - symptomatic VT

Deanfield et al. Br Heart J 1980;44:248–253.
Silka et al. J Am Coll Cardiol 1998;32:245–251.

196 TOF repair
F/U 1-20 yr (mean 10yr)

Table 1 *Conduction defects on preoperative, postoperative, and follow-up electrocardiograms*

Conduction pattern	*Preoperative*	*Postoperative*	*Follow-up*
	No. of patients (%)	*No. of patients (%)*	*No. of patients (%)*
Normal	170 (87)	7 (4)	2 (1)
Incomplete RBBB	22 (11)	2 (1)	2 (1)
Complete RBBB	4 (2)	187* (95)	192 (98)
RBBB + LAD	0 (0)	17 (9)	33 (17)

RBBB, right bundle-branch block; LAD, left axis deviation.
* Includes the 17 patients who also had left axis deviation.

Table 2 *Frequency of perioperative arrhythmias related to electrocardiographic conduction defects*

Conduction defect	*Total patients*	*First degree AV block*	*Complete heart block*	*Nodal rhythm*	*Ventricular extrasystoles*
Total group	196	18 (9%)	9 (5%)	10 (5%)	21 (11%)
RBBB	158	12 (8%)	7 (4%)	9 (6%)	17 (11%)
RBBB+LAD	17	4 (23%)	1 (6%)	0 (0%)	3 (18%)
Progressive conduction defect	21	2 (10%)	1 (5%)	1 (5%)	1 (5%)

LAD, left axis deviation; AV, atrioventricular.

Deanfield et al. Br Heart J 1980;44:248–253.

Risk of sudden cardiac death after CHD repair

- population based prospective study
- Oregon 1958~1996, < 19yr
- 3,589 total, 41 SCD

Table 1. Specific Congenital Heart Defects and Incidence of Sudden and Nonsudden Cardiac Death

	No. (%) of Pts With Complete Follow-Up	Total Follow-Up (pt-yr)	Sudden Cardiac Death		Nonsudden Cardiac Death	
			No.	Incidence/ 1,000 Pt-yr	No.	Incidence/ 1,000 Pt-yr
ASD	622 (86%)	7,904	0	0	0	0
VSD	527 (87%)	6,354	1	0.2	8	1.2
AVSD	254 (87%)	2,217	2	0.9	15	6.7
PDA	623 (82%)	8,753	0	0	4	0.4
PS	241 (91%)	3,568	1	0.3	2	0.6
AS	169 (94%)	1,860	10	5.4	9	4.8
CoA	536 (92%)	6,706	9	1.3	17	2.5
TOF	445 (91%)	7,082	11	1.5	9	1.3
D-TGA	172 (95%)	1,413	7	4.9	10	6.9
Total	3,589	45,857	41	0.9	74	1.6

AS = aortic stenosis; ASD = atrial septal defect; AVSD = atrioventricular septal defect; CoA = coarctation of the aorta; PDA = patent ductus arteriosus; PS = pulmonary stenosis; pt-yr = patient-years; Pts = patients; D-TGA = dextro-transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

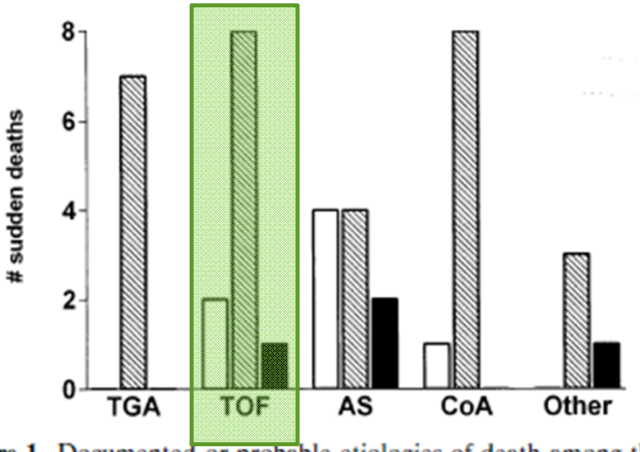


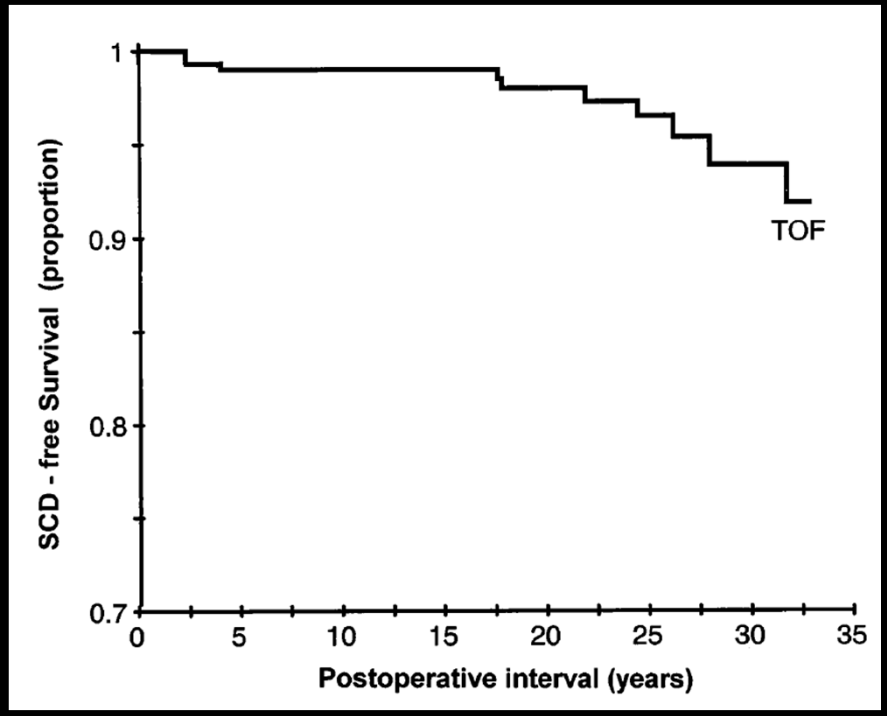
Figure 1. Documented or probable etiologies of death among the 41 patients who died suddenly (arrhythmia **[hatched bars]**, circulatory **[open bars]**, congestive heart failure **[solid bars]**). The anatomic diagnoses of the other patients were pulmonary stenosis in one patient, ventricular septal defect in one and AV septal defect in two.

Silka et al. J Am Coll Cardiol 1998;32:245–251.

Recent studies : sudden death is considerably lower

Silka et al. risk of SCD

- 1.2% at 10 years
- 2.2% at 20years
- 4% at 25 years
- 6% at 30 years



Silka et al. J Am Coll Cardiol 1998;32:245–251.

Risk group of ventricular arrhythmia & SCD

- ventricular arrhythmias : m.c. in older preOp patients

  -> remain after repair

Table 3 *Arrhythmias and conduction defects in patients who died suddenly*

Case no.	*Postoperative conduction defect*	*Follow-up conduction defect*	*Postoperative arrhythmia*	*Late arrhythmia*
1	RBBB	RBBB	—	—
2	RBBB	RBBB	—	—
3	RBBB	RBBB	—	Supraventricular tachycardia
4	RBBB	RBBB	—	—
5	RBBB	RBBB + LAD	—	—
6	RBBB	RBBB	—	Ventricular extrasystoles
7*	RBBB	RBBB	—	—
8	RBBB + LAD	RBBB + LAD	—	—
9	Normal	RBBB	Junctional tachycardia	—

LAD, left axis deviation; RBBB, right bundle-branch block.

*Operation performed at The Hospital for Sick Children, Great Ormond Street.

Postoperative arrhythmias

a/w : only surgery age

not postOp status, duration of F/U, hemodynamic status.

Deanfield et al. Br Heart J 1980;44:248–253.

- predictor of sudden death in 125 adult TOF
  - PR degree
  - Hx of sustained VT
  - QRS duration
    - > 180ms
  - LV dysfunction

Table 3. Clinical Outcomes, Electrocardiographic and Hemodynamic Data for the SCD and Control Groups

	SCD Group (n = 12)	Control Group (n = 125)	p Value
Symptoms			
Dyspnea	7 (58%)	31 (25%)	0.03
Orthopnea	5 (42%)	15 (12%)	0.01
Palpitations	10 (83%)	41 (33%)	0.01
Presyncope or syncope	7 (58%)	14 (11%)	< 0.01
Edema	5 (42%)	16 (13%)	0.01
NYHA class ≥ 2	4 (33%)	18 (14%)	0.12
Atrial fibrillation or atrial flutter	3 (25%)	23 (19%)	0.74
Ventricular tachycardia	5 (42%)	7 (6%)	< 0.01
Medications			
Anticoagulation/antiplatelets	7 (58%)	29 (23%)	0.05
Antiarrhythmic drugs	8 (67%)	18 (14%)	< 0.01
ACE inhibitors	5 (42%)	16 (13%)	0.02
Diuretics	6 (50%)	13 (10%)	< 0.01
Beta-blockers	2 (17%)	5 (4%)	0.05
ECG data			
Number of patients with data	9	123	—
Mean (ms) QRS duration	170 ± 14	153 ± 21	0.02
QRS ≥ 180 ms	5 (56%)	16 (13%)	0.02
Echocardiographic data			
Mean age (yrs) of last echocardiogram	43 ± 18	38 ± 14	0.58
Moderate or severe LV dysfunction	5 (42%)	11 (9%)	< 0.01
RV > mild dilation	7 (54%)	40 (32%)	0.09
Mean RVSP (mm Hg)*	46 ± 19	44 ± 13	0.54
PR ≥ moderate	11 (92%)	64 (51%)	0.02
TR ≥ moderate	6 (50%)	38 (30%)	0.38
MR ≥ moderate	2 (17%)	7 (6%)	0.06
AR ≥ moderate	0 (0%)	1 (1%)	0.87

- 59 TOF pt (18 ±5 yr)
  ↑ severity of PR & RV dilation
  higher risk or ventricular arrhythmias.

  Zahka et al. Circulation 1988;78(pt 2):14–19.

- multi center study (800 TOF)
  significant PR
  a/w ventricular tachycardia & higher risk of sudden death

  Gatzoulis et al. Lancet 2000;356:975–981.

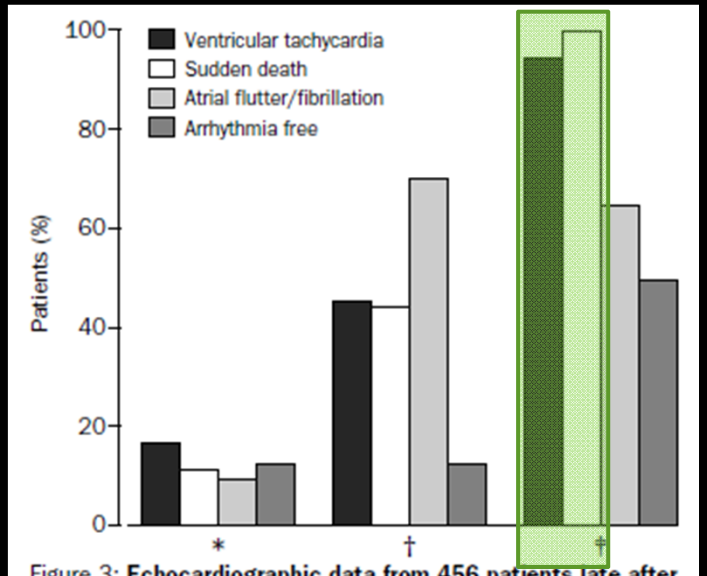


Figure 3: **Echocardiographic data from 456 patients late after repair of tetralogy of Fallot**
Data obtained during preceding 12 months from final patient assessment in 1995, or before occurrence of arrhythmia and sudden death.
*Right-ventricular systolic pressure (>60 mm Hg). †Tricuspid regurgitation ≥moderate). ‡Pulmonary regurgitation (≥moderate)

Floppy Mitral Valve/ Mitral Valve Prolapse

FMV/MVP : almost normal ECG

- Nonspecific ST- & T- changes & T-wave inversion
- Ventricular arrhythmia d/t
  - Autonomic dysfunction
  - Papillary muscle traction/ventricular stretch
  - WPW syndrome
  - Long QT syndrome
  - Mechanical stimulation of myocardium by leaflets
  - Abnormal innervation of floppy mitral valve
  - Endocardial friction lesions
  - Platelet aggregation; fibrin deposits
  - Myocardial fibrosis

103 children with FMV/MVP vs 50 normal children

- FMV/MVP
  - 16% exercise-induced premature ventricular beats (PVBs)
  - 38% PVBs on ambulatory monitoring
  - VT 4% during exercise, 8% on ambulatory monitoring.
- Control
  - none single PVB in exercise
  - 8% uniform PVBs on ambulatory monitoring

Kavey et al. J Pediatr 1984;105:885–890.

Table II. Ventricular premature complexes on treadmill exercise and on 24-hour ambulatory ECG in children with mitral valve prolapse and in controls

	Mitral valve prolapse (n = 103)		*Control (n = 50)*	
	Treadmill exercise	*Ambulatory ECG*	*Treadmill exercise*	*Ambulatory ECG*
No VPCs	87	64	50	46
Grade 1	4	24	0	4
Grade 2	8	7	0	0
Grade 3	2	5	0	0
Grade 4	2	3	0	0
Supraventricular tachycardia	2	3	0	0

108 MVP children vs 70 normal controls

- Benign arrhythmias on 24-hour Holter monitoring
  - MVP 49.1%
  - control 21.4%
  - PVBs : m.c.
- Isolated PVBs
  - FMV/MVP 27.3% vs controls (8.5%)

Ohara N et al. Acta Paediatr Jpn 1991;33:467-475

Isolated atrial premature beats

- 23.6% of MVP children vs 10% in the controls

Boudoulas H, Wooley CF, eds. The Floppy Mitral Valve, Mitral Valve Prolapse, and Mitral Valvular Regurgitation, 2000.

D-TGA(Transposition of great arteries)

In newborn period

dysrhythmias are rare

24-hour ambulatory ECG

-> short bradycardia & junctional rhythm may occur

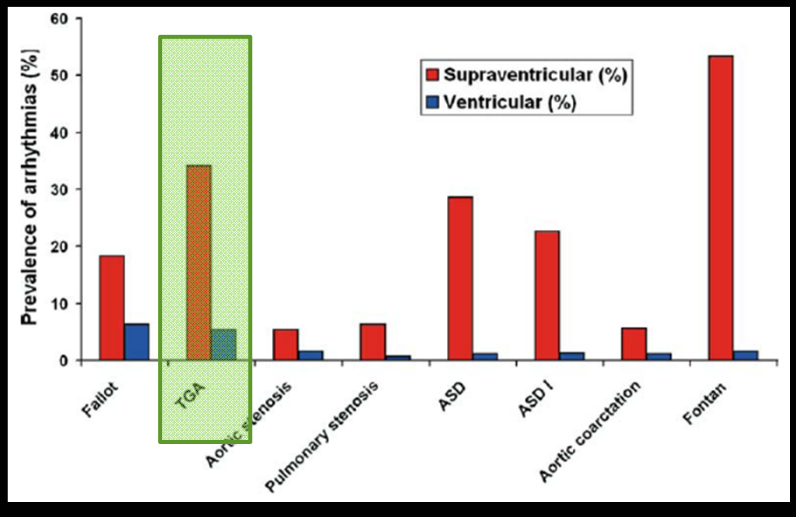
In a large Dutch registry

d-TGA/atrial switch

-35% atrial arrhythmias (second only to single-ventricle)

-7% ventricular arrhythmias

(second only to TOF)

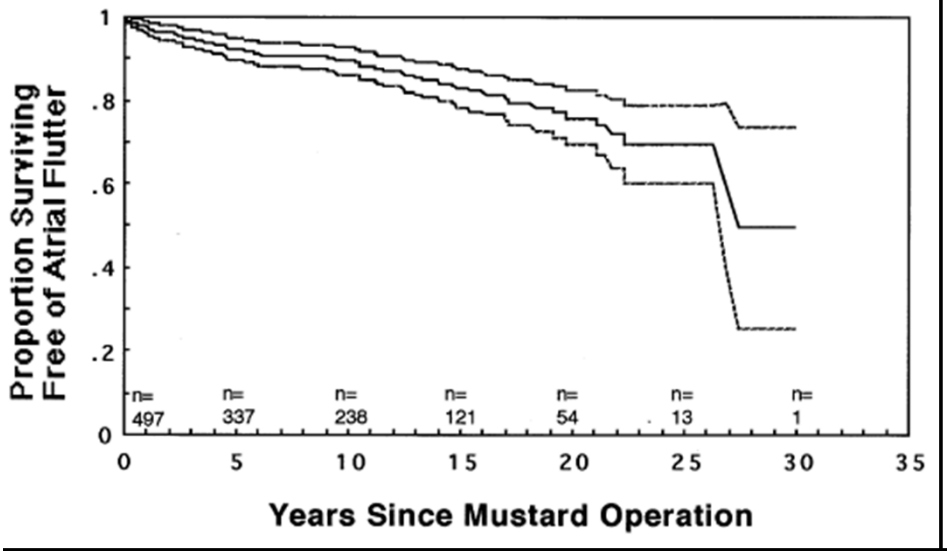


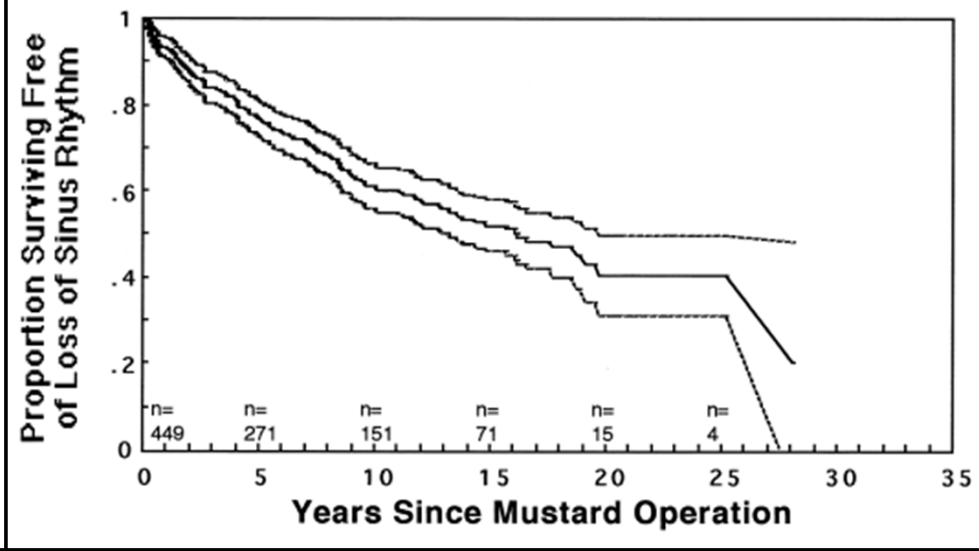
Vander Velde ET et al. Eur J Epidemiol 2005;20;549~557

Atrial switch (Mustard)

Atrial flutter 14%

Sinus rhythm : only 40% after 20 years





Gelatt M et al. J Am Coll Cardiol 1997;29:194~201

Atrial switch (Mustard vs Senning)

losing sinus rhythm over time

only 18% / 15 years post repair

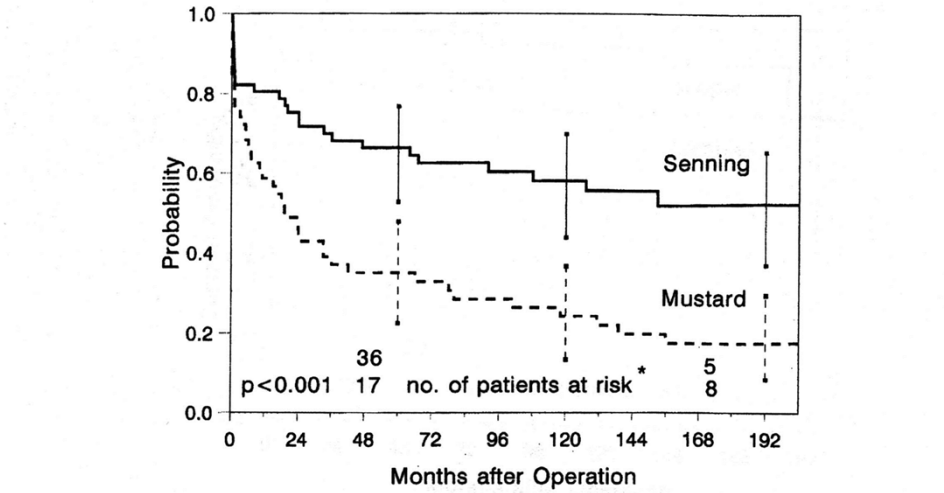
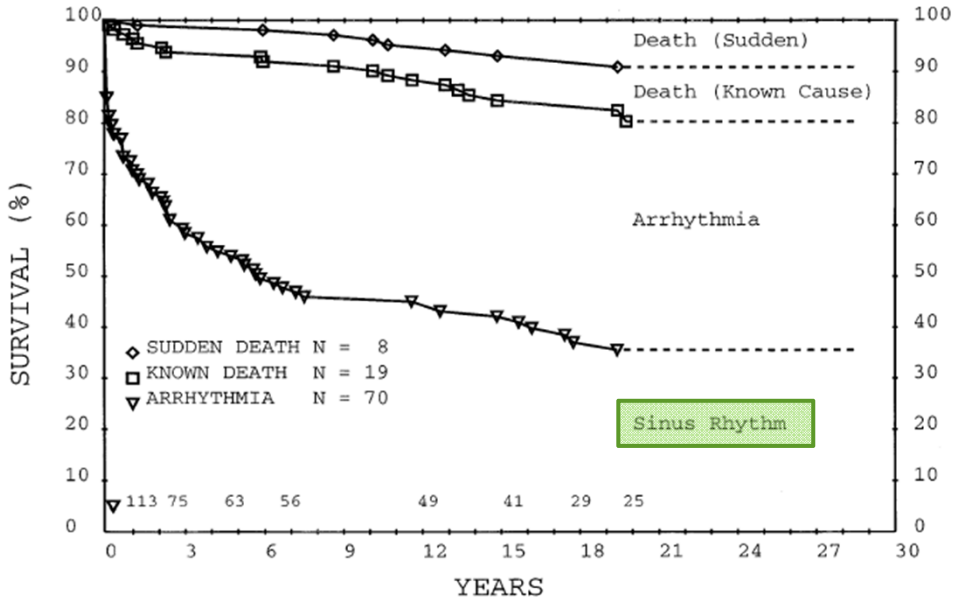


Fig. 1. Kaplan-Meier curves for survival in the absence of rhythm disturbances of hospital survivors of atrial correction for transposition of the great arteries. (*Unbroken lines*, Senning operation; *broken lines*, Mustard operation.) The p value for comparison between rates in the Senning and Mustard patients was calculated with the log-rank test. *Vertical lines* represent 95% confidence intervals at 5-, 10-, and 16-year follow-up. *See Statistics section for method of censoring.



Mustard vs Senning

Helbing WA et al.
J Thorac Cardiovasc Surg 1994;108:363~372

Mustard

Wilson NJ et al.
J Am Coll Cardiol 1998;32:758-765

Sudden cardiac death in post–atrial switch d-TGA most important unpredictable event

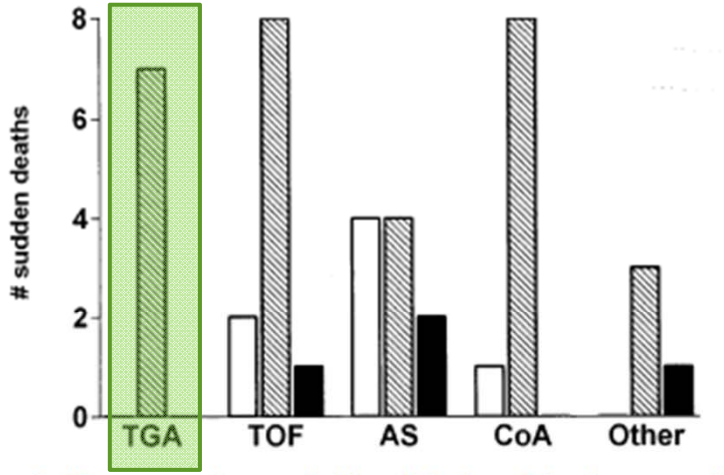


Figure 1. Documented or probable etiologies of death among the 41 patients who died suddenly (arrhythmia **[hatched bars]**, circulatory **[open bars]**, congestive heart failure **[solid bars]**). The anatomic diagnoses of the other patients were pulmonary stenosis in one patient, ventricular septal defect in one and AV septal defect in two.

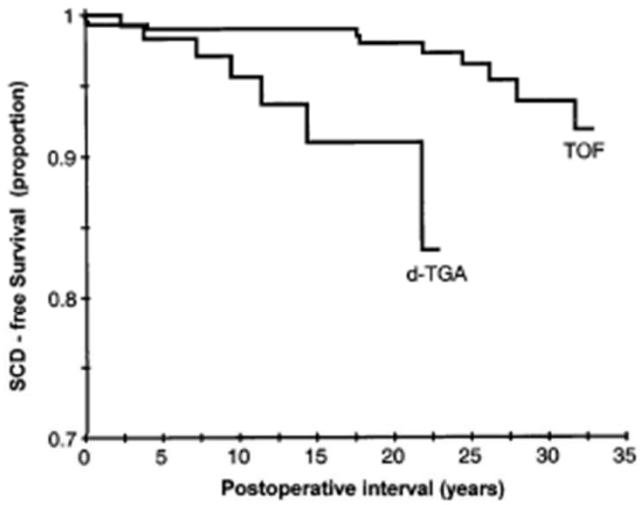


Figure 2. Actuarial probability of SCD-free survival after surgical treatment of cyanotic congenital heart defects.

Silka MJ et al. J Am Coll Cardiol 1998;32:245~251

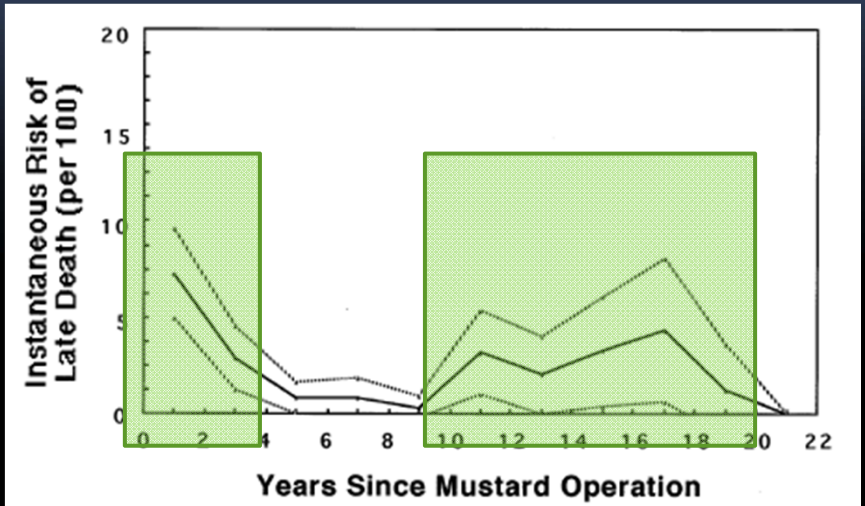


Figure 2. Instantaneous hazard (risk) of late death for each 2-year period after the Mustard procedure. Top and bottom lines indicate 95% confidence intervals.

Gelatt M et al. J Am Coll Cardiol 1997;29:194~201

SCD related to atrial or ventricular arrhythmias

atrial arrhythmias leading to SCD(arrhythmia Sx, AFL/AF)

Table 2. Variables for Which Conditional Logistic Regression Analysis Was Performed, Noted in Mean OR With 95% CI

	p Value	OR (95% CI)
Symptoms		
Symptomatic*	<0.0005	6.45 (2.42–17.24)
Arrhythmic symptoms*	0.003	21.60 (2.80–166.79)
Heart failure symptoms*	0.001	4.44 (1.85–10.62)
ECG		
QRS duration	0.723	0.32 (0.001–175.66)
QT interval	0.668	0.16 (0.000–734.27)
QTc interval	0.193	1084.50 (0.029–4.1E + 07)
QRS duration >100 ms	0.251	1.980 (0.618–6.324)
QT dispersion	0.126	0.988 (0.973–1.003)
Heart rate	0.054	1.017 (1.000–1.035)
Basal heart rhythm nonsinus	0.790	1.112 (0.509–2.427)
Chest X-ray		
Enlarged heart size	0.053	2.227 (0.989–5.000)
24-h Holter		
Basal heart rhythm nonsinus	0.037	5.260 (1.10–25.00)
Documented episodes of arrhythmia	0.431	1.770 (0.44–7.25)
Mean heart rate	0.527	0.980 (0.919–1.044)
Minimum heart rate	0.952	1.001 (0.956–1.050)
Maximum heart rate	0.803	0.996 (0.965–1.028)
History of arrhythmia in follow-up		
Documented arrhythmia*	0.005	3.473 (1.451–8.310)
Documented SND in follow-up	0.035	2.405 (1.065–5.432)
Documented AFL/AF in follow-up*	0.001	4.866 (1.900–12.462)
Arrhythmia treatment		
Pacemaker implantation	0.550	0.641 (0.149–2.758)
Medication treatment*	0.002	5.159 (1.863–14.283)

*Statistically significant risk factor ($p < 0.005$).
AFL/AF = atrial flutter/atrial fibrillation; CI = confidence interval; OR = odds ratio; SND = sinus node disease.

Kammeraad JA et al. J Am Coll Cardiol 2004;44:1095-1102

Corrected Transposition of the Great Arteries

Ventricular inversion

-> ventricular bundle branches : also inverted

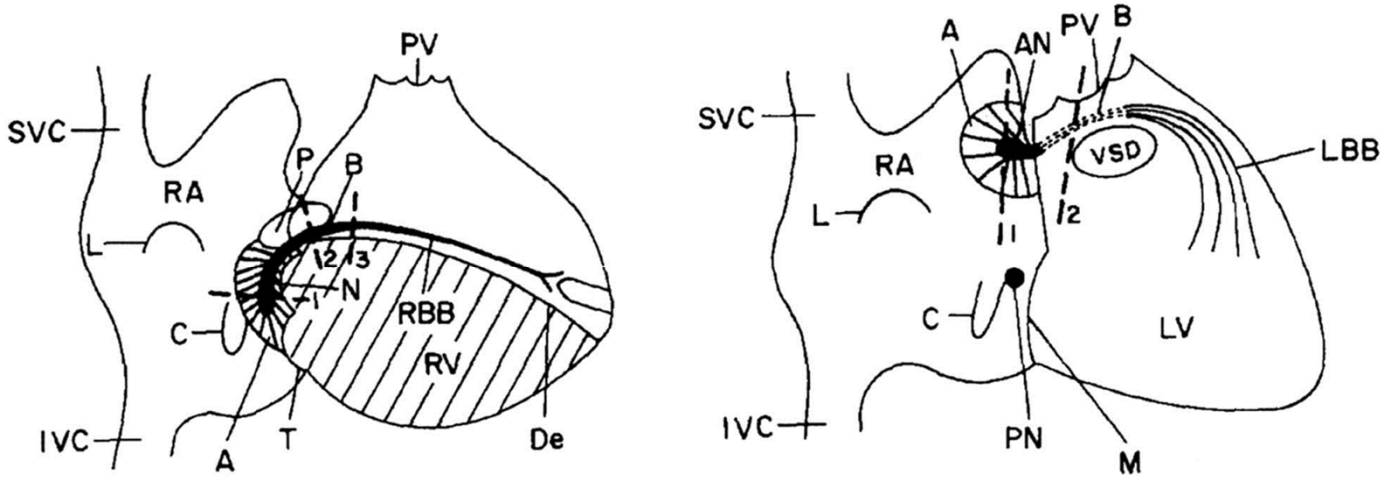


FIGURE 7. Diagrammatic sketches of the conduction system in pure levocardia **(left)** (atrioventricular [A-V] concordance [*d*-loop]) and in one type of mixed levocardia **(right)** (atrioventricular discordance [*l*-loop] as seen from the right side, and the sites of interruption that may be present. Note that the bundle of His in mixed levocardia is depicted by **dotted lines** to indicate that it is coursing on the parietal wall to reach the septum. **Dashed lines** labeled 1, 2 and 3 are areas in which interruption of the conduction system has been recorded in congenital A-V block: 1 = interruption in the A-V node and its approaches; 2 = interruption in the bundle of His; and 3 = interruption in the bundle branches. A = approaches to the A-V node; AN = anterior A-V node; B = bundle of His; C = opening of coronary sinus; De = demarcation between sinus and infundibulum of right ventricle; IVC = inferior vena cava; L = limbus fossae ovalis; LBB = left bundle branch; LV = left ventricle; M = mitral valve; N = normal (posterior) A-V node; P = pars membranacea; PN = posterior A-V node; PV = pulmonic valve; RA = right atrium; RBB = right bundle branch; RV = right ventricle; SVC = superior vena cava; T = tricuspid valve; VSD = ventricular septal defect.

Bharati S et al. Am J Cardiol 1978;42:147-153

Complete heart block
congenital and post surgical

Table 1
Incidence of conduction abnormalities associated with L-TGA[a]

Source	Patient population	Heart block incidence	Atrial arrhythmias	Pacemakers
Bjarke et al. [5]	*n* = 101 43% single ventricles	*n* = 17 (17%) 1st: 7 (7%) 2nd: 2 (2%) 3rd: 8 (8%)	*n* = 12 (12%) (pre-surgical)	
Daliento et al. [7]	*n* = 17 All two ventricles	*n* = 5 (29%)		
Friedberg et al. [4]	*n* = 60 42% SV 52% VSD 6% IVS	*n* = 19 (32%) 1st: 10 (17%) 2nd: 2 (3%) 3rd: 7 (12%)		
Huhta et al. [3]	*n* = 107 All two ventricle 77% VSD	*n* = 23 (22%) CCHB: 4 (4%)	*n* = 4 (4%) WPW: 2 AFL: 2	*n* = 9 (39%)
Lundstrom et al. [20]	*n* = 111 All two ventricle	*n* = 35 (32%) CCHB: 4 (4%) Spont: 17 (15%) SxHB: 14 (13%)		*n* = 9 (8%) All for CHB
Fyler [21] (includes subset of Friedberg et al. population)	*n* = 90 All two ventricle	*n* = 29 (32%)		
Michigan Congenital Heart, 1998	*n* = 126 SV = 57 (45%)	*n* = 40 (32%) 1st: 11 (9%) 2nd: 4 (3%) 3rd: 19 (15%) – CCHB 6 SxHB: 6 (5%)	*n* = 19 (15%) WPW: 5 AFL: 6 AF: 1 PSVT: 2 SSS: 5	*n* = 27 (21%) SSS: 5 (4%) 2nd HB: 1 (1%) CHB: 10 (8%) SxHB: 9 (7%)

Fischbach P et al. Prog Pediatric Cardiol 1999;10:37-43

Complete heart block

10% at initial presentation

Bharati S et al. Am J Cardiol 1978;42:147-153

↑ 2%/year during follow-up

Daliento L et al. Am J Cardiol 1986;58:314-318

50% : pacemaker therapy

Atrial arrhythmias

38%

atrial fibrillation, atrial flutter, and SVT

→Adult patients

routine exams, ECG, 24-hour ambulatory monitoring

Hypoplastic Left Heart Syndrome

Arrhythmias after Fontan procedure

late atrial arrhythmias : 10- 45%

d/t sinus node dysfunction (atrial suture lines)

↑ atrial pressure

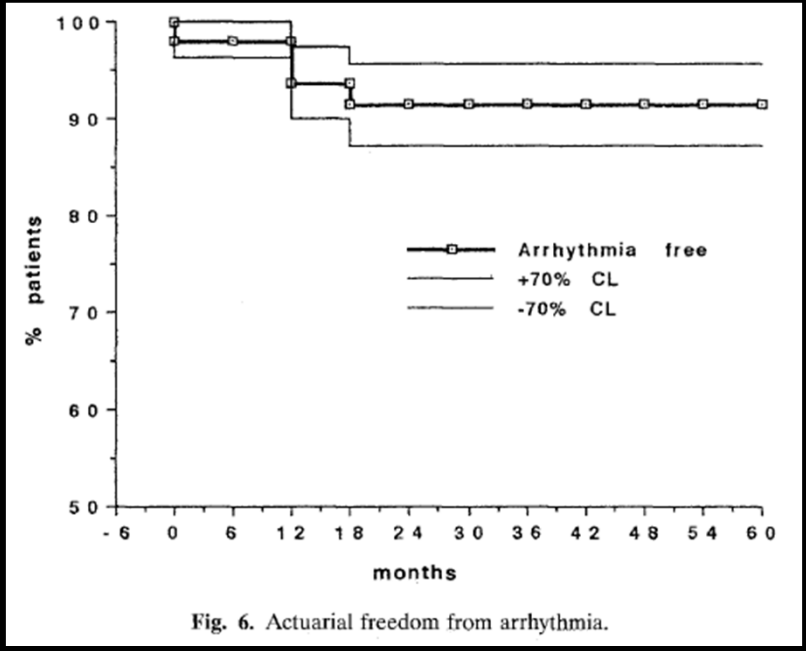


Fig. 6. Actuarial freedom from arrhythmia.

Amodeo A, et al. J Thorac Cardiovasc Surg 1997;114:1020–1030.

TABLE 8. Early nonfatal morbidity

Complication	n	% of total
Prolonged pleural effusion	36	30
Atrial arrhythmias	25	21
Early reoperation	13	11
Bleeding	11	9
Other	2	1.7
Permanent pacemaker	8	7
Pneumonia	6	5
Renal failure	3	2.5
Cardiac arrest (surviving)	3	2.5
Tamponade	2	1.7
Cerebral vascular accident	2	1.7
Tracheostomy	1	0.8
Sternal wound infection	1	0.8
Gastrointestinal hemorrhage	1	0.8
Colitis	1	0.8

Burkhart HM et al. J Thorac Cardiovasc Surg 2003;125:1252-1258

TABLE 11. Arrhythmias in 215 Survivors

Results from follow-up questionnaire	5 Years postop Patients No.	%	Currently Patients No.	%
Syncope	18	8	17	8
Rapid heart rate (tachycardia)	44	20	45	21
Slow heart rate (bradycardia)	17	8	15	7
Palpitations	51	24	60	28
Atrial flutter or fibrillation	26	12	41	19
Premature ventricular contractions	13	6	15	7
Ventricular tachycardia	9	4	13	6
Pacemaker	†	†	22	10
Number of antiarrhythmic medications*				
0	179	83	167	78
1	31	14	40	19
2	5	2	8	4

*Excluding digitalis.
†Presence of a pacemaker was only asked for patient's current status.

Driscoll D et al. Circulation 1992;85:469-496

Extracardiac Fontan

for reducing the incidence of atrial arrhythmias

by minimizes atrial suture lines

lessens the atrial hypertension

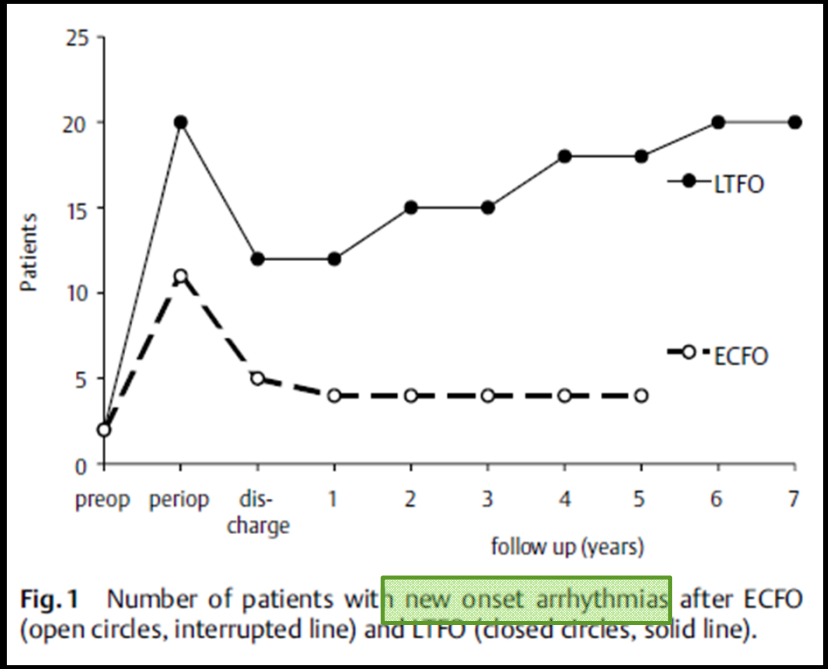


Fig. 1 Number of patients with new onset arrhythmias after ECFO (open circles, interrupted line) and LTFO (closed circles, solid line).

Ovroutski S, et al. Thorac Cardiovasc Surg 2001;49:334–337.

Double Outlet Right Ventricle

AV node
- normal posterior position

conduction system
- penetrated the right side of the central fibrous body

AV bundle
- lie in the inferior wall of the defect
- passed beneath the crest of the ventricular septum

Stellin G, et al. J Thorac Cardiovasc Surg 1991;102:849–855.

First-degree AV conduction delay : common

Double Inlet Left Ventricle

Conduction abnormalities similar : corrected TGA

AV node

anterolaterally at acute margin of the right AV valve orifice

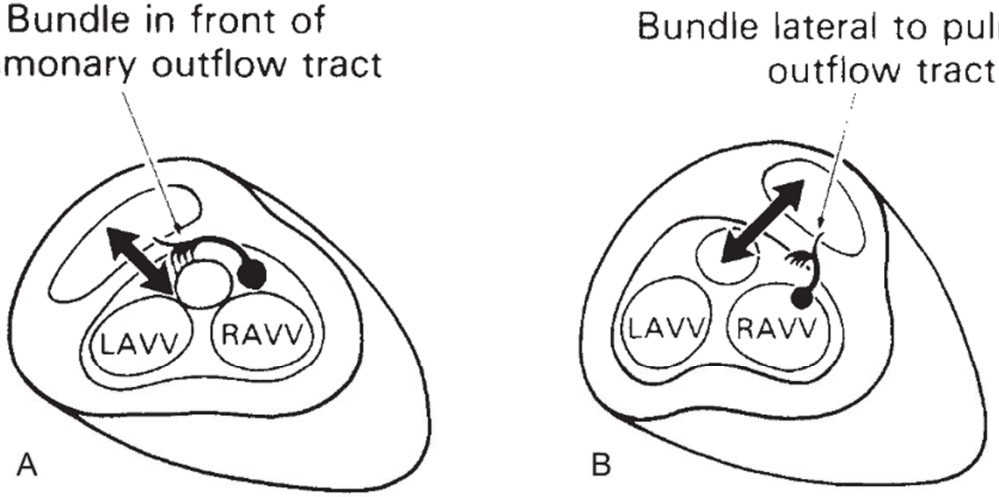


FIGURE 54.8 Conduction tissue. Schematic illustration of the course of the nonbranching bundle of the conduction tissue along the right rim of the ventricular septal defect in double-inlet left ventricle (DILV). **A:** The findings in DILV with a left-anterior subaortic right ventricle. **B:** The conduction tissue in DILV with a right-anterior subaortic right ventricle. LAVV, left atrioventricular valve; RAVV, right atrioventricular valve. (From Davies MJ, Anderson RH, Becker AE. Atrioventricular conduction tissues in congenital heart disease. In: Davies MJ, Anderson RH, Becker AE, eds. *The Conduction System of the Heart*. London: Butterworth, 1983:137, with permission.)

Allen et al. Moss and Adams' heart disease in infants, children, and adolescents. 2008.

merism

us node (SN)

by atrial appendages

-right isomerism : bilateral SN

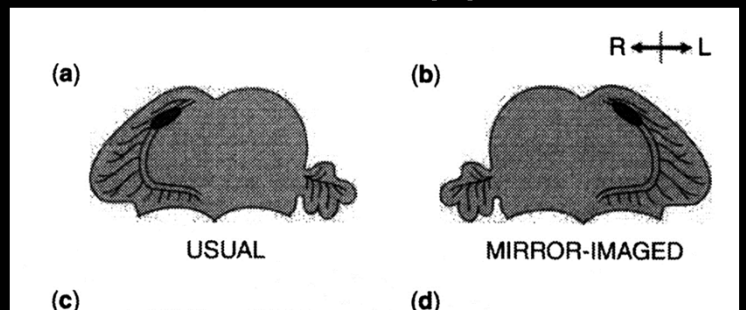
terminal crest & cavoatrial junction in normal fashion

22. Smith A., et al. Cardiol Young 2006; 16:437-454.

-left isomerism

no terminal crests, and no RA appendages.

SN cannot occupy its normal position

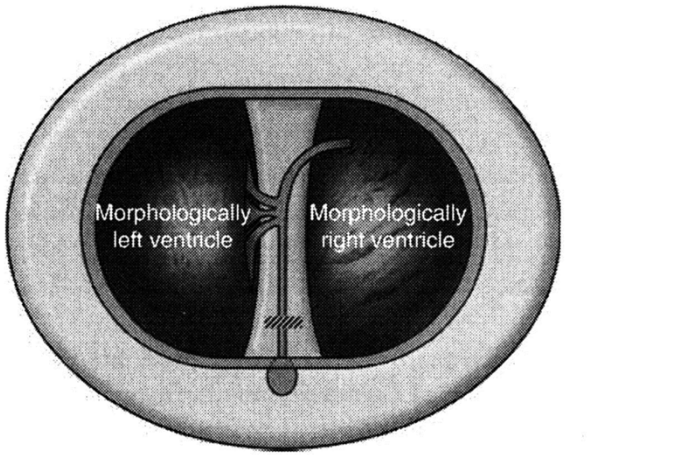


22. Smith A., et al.

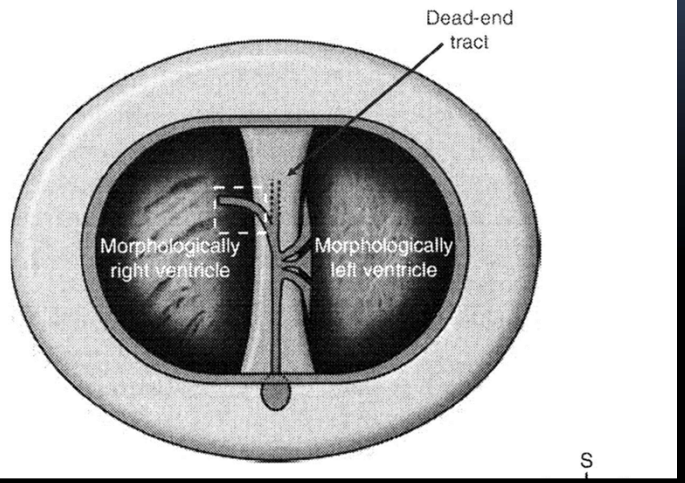
Cardiol Young 2006; 16:437-454.

ventricular AV connection
AV node : regular position
If right hand topology
AV bundle - postero-inferior penetrating
axis of conduction tissue - deviated posteriorly in AVSD
If left hand topology
- anterior AV node + conduction of cc TGA
or- sling of conduction tissue -> 2 AV nodes

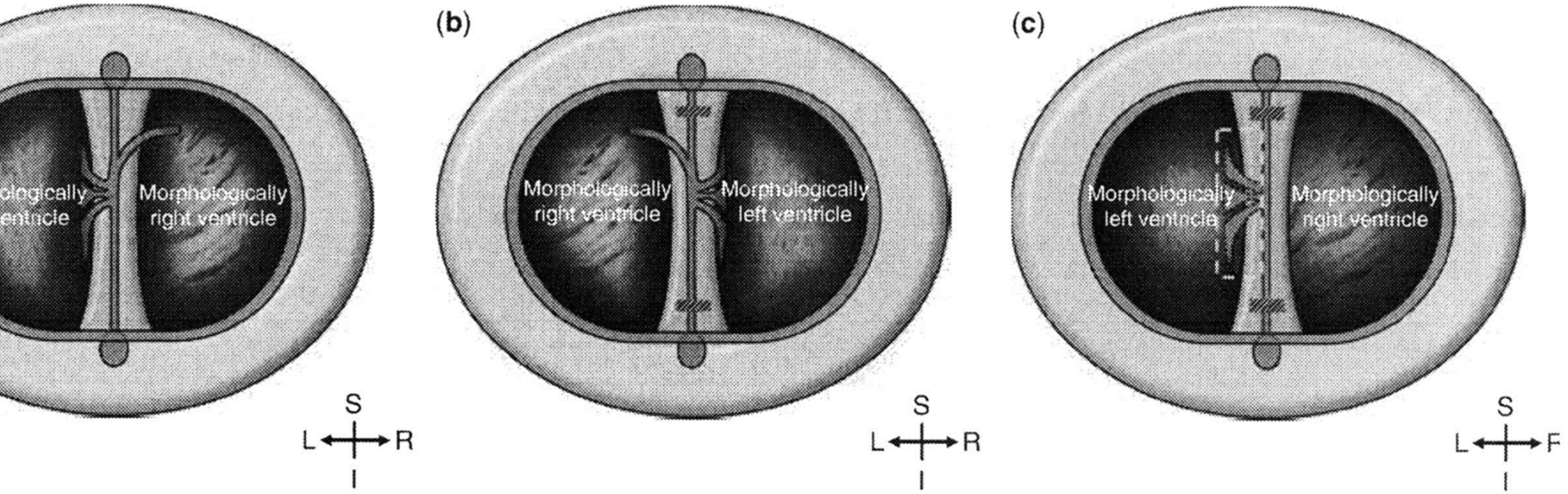
iventricular AV connection
LV dominant : AV node – ant. position
bundle vary
RV dominant
rud. LV in left – conduction sys. normal
rud. LV in right- ant. node. or sling
litary and indeterminate ventricle



BV connection,
Rt. topology
Lt. isomerism



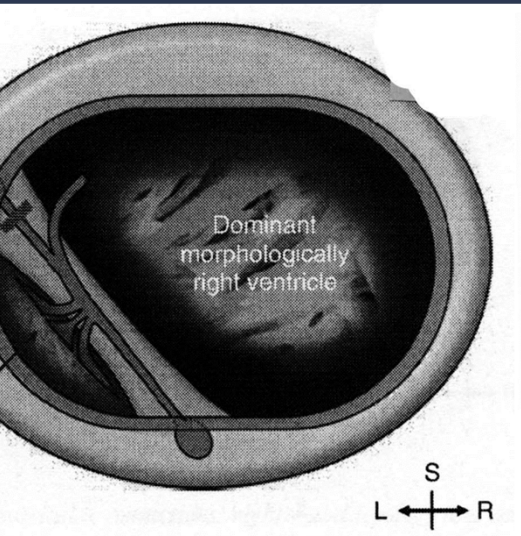
BV connection,
Lt. topology
Lt. isomerism



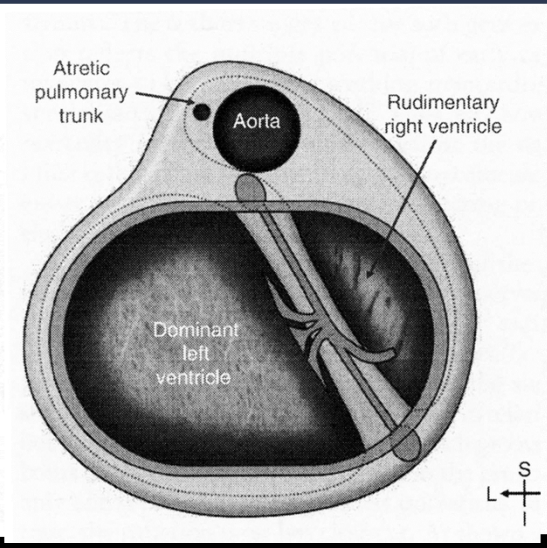
V connection,
t. topology
t. isomerism
VSD

BV connection,
Lt. topology
Lt. isomerism
AVSD

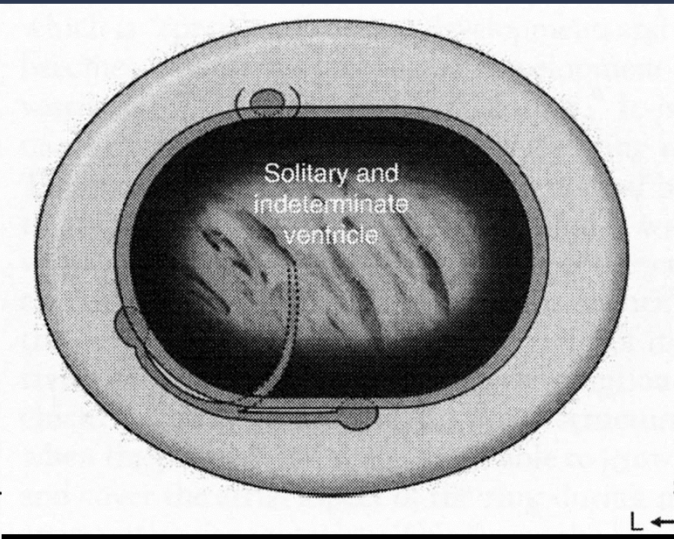
BV connection,
Rt. topology
Rt. isomerism
AVSD



IRV,
t./Rt. isomerism



DILV,
Rt. Isomerism
CAVV



DI,
Indeterminate V
Rt. isomerism

rhythmias after Cardiopulmonary bypass

ter CPB

- 25% : arrhythmias occur

k factors

- longstanding volume overload
- ventricular hypertrophy
- myocardial ischemia
- ventriculotomy multiple suture lines
- electrolyte disturbances.

hythmias

- occur < 48 hrs of surgery
- bradyarrhythmias- sinus bradycardia, complete block
- tachyarrhythmias - SVT, atrial flutter, atrial fibrillation

T : common after pediatric cardiac surgery
young infants after repair of TOF
closure of VSD & AVSD

atients Who Experienced Junctional Ectopic Tachycardia

Diagnosis	Surgical Procedure	Postoperative Day for Initial Occurrence of JET (number of days of JET total)
	Repair	1 (1)
	Repair	1 (1)
	Repair	1 (1)
	Repair	1 (1)
PV	Repair	2 (1)
A	RV-PA conduit	1 (1)
A s/p ocalization	Residual VSD closure	4 (1)
AVC	Repair	4 (1)
SD/CoA	Repair/ECMO	1 (1)
SD/CoA	Repair	3 (1)
ventricle (H)	Stage 1	1 (1)
ventricle (H)	Stage 1	2 (1)
ventricle (H)	Stage 1	1 (1)
ventricle (H)	BTS	3 (1)
ventricle	Hemi-Fontan/SVC plasty	2 (3)
ventricle	Extracardiac Fontan	5 (1)
ventricle	Extracardiac Fontan	1 (1)
ventricle	Extracardiac Fontan	4 (4)
C	Repair	1 (1)
C	Repair	1 (1)
C	Repair	1 (3)
C	Repair	1 (1)
R/CDH	Repair	2 (2)
oA	Repair	1 (1)
oA	Repair	1 (1)
S	Repair	1 (2)

Table 1. Success Rate Versus Anatomy and Tachyarrhythmia Type

Cardiac Anatomy (no.)	VT	VT/VF	AutoAT	IART	JET	Other SVT	Total
Postop Fontan (8)				1/2	5/5	0/1	6/8
Postop VSD or AVC (6)		0/1			4/5		4/6
Myocarditis (4)	1/2		2/2				3/4
Hamartoma/rhabdo (4)	3/3					1/1	4/4
Postop TAPVR (3)			2/2		1/1		3/3
Postop arterial switch (2)				1/1	1/1		2/2
Postop truncus (2)		1/1		1/1			2/2
Postop tetralogy (2)				1/1	1/1		2/2
Hypertrophy CM (2)		1/1	1/1				2/2
Dilated CM (2)	1/1	0/1					1/2
Normal (2)					1/1	0/1	1/2
Others (3)	1/1	0/1	1/1				2/3
Total (40)	6/7	2/5	6/6	4/5	13/14	1/3	32/40

Data presented are number of patients with successful treatment/total patients treated. AutoAT = automatic atrial tachycardia; AVC = atrioventricular canal defects; CM = cardiomyopathy; IART = intraatrial reentrant tachycardia; JET = junctional ectopic tachycardia; Postop = postoperative; rhabdo = rhabdomyoma; SVT = supraventricular tachycardia; TAPVR = total anomalous pulmonary venous return; VF = ventricular fibrillation; VSD = ventricular septal defect; VT = ventricular tachycardia.

Perry JC et al. J Am Coll Cardiol 1996;27:1246-1250

nclusion

ongenital heart diseases
conduction disturbance occur
-congenital
-acquired : hemodynamic effect on conduction system
surgery related

nduction disturbance in CHD
understanding
Identification of risk group
minimize surgery related conduction dis.
regular evaluation – ECG, 24hr holter, EPS
early intervention