Acquired conduction disturbance in structurally normal heart in children

Mi Young Han, M.D., PhD. Kyung Hee University Hospital



Incidence

- Cardiac conduction disturbances detected in a pediatric population
 - 432,166 elementary and high school students
 - Prevalence of CCD increased with age :
 0.48% → 0.97%
 - higher in males
 - Incomplete RBBB(0.32%), complete RBBB(0.11%), VPC(0.11%), WPW syndrome(0.067%)
 - Sensitivity of IRBBiBeing Screen Pring; 159185-89
 ASD was 34 67%

Classification

- First AV Block
- Second AV Block
- Complete AV Block
- Intraventricular block
- Drug-induced long QT syndrome

Causes of acquired heart block

- medications
- myocardial inflammation
- Myopathy
- infection (Lyme disease, viral myocarditis, endocarditis)
- Hypothyroidism
- surgical trauma
- high levels of vagal tone
- Anorexia nervosa

Drug-induced atrioventricular block

- AV block (diagnosed during therapy with antiarrhythmic medication) that resolved when the drug discontinued and never recurred during a follow-up period (≤ 3wks)
- Beta-blockers, nondihydropyridine calcium channel antagonists(verapamil, diltiazem)

Drug-induced atrioventricular block : prognosis : benign ?



Infectious myocarditis - infectious

ad	<u>pnt</u>		
	Bacteria	Virus	Parasites
	Diphtheria	Epstein-Barr virus	Candidosis
	Cholera	Mumps	Aspergillosis
	Leptospirosis	Cytomegalovirus	Trichinosis
	Mycoplasma	Rubella	Hydatidosis
	pneumoniae		
	Rickettsia	Poliomyelitis	Toxoplasmosis
	Streptococcus	Parainfluenza	Paludism
	Meningococcus	Influenza	Leishmaniosis
	Listeriosis	Varicella	Bilharziosis
	Staphylococcus	Herpes	Trypanosoma gambiense
	Tuberculosis	measles	Cysticercosis
	Syphilis	Arbovirosis	Opisthorchiasis
	Shigella	Hepatitis	Paragonimiasis
	Salmonella	HIV	
		Coxsackie B	
		Adenovirus	
		Respiratory syncytial virus	
		Hantaan virus	
		Enterovirus	

The underlined data are agents for which the implantation of a permanent pacemaker has been reported. (complete references available on request).

Maury et al., J of Electrophysiology 2008:41;665-667,

Infectious myocarditis

 Acquired AV block in children (even without myocardial systolic dysfunction)→ r/o myocarditis

Biopsy

- Acute phase : injury of the conduction system, with mitochondrial inclusions, lymphocytes, mononuclear cells infiltrates, areas of inflammation and necrosis
- Delayed phase : scar fibrosis Batra et al. Pediatr Cardiol 2003;24:495-497

Infectious myocarditis

- Electrophysiologic investigations
 - Supra-hisian, intra-hisian, or infrahisian block
 - Distal lesion : intraventraventricular block
- Treatment
 - Transient pacing (2/3; recovered at 1 week)
 - Permanent pacemaker
 - 20-30% of the cases Batra et al. Pediatr Cardiol 2003;24:495-497

Infectious myocarditis

- Clinical course of CHB a/w acute myocarditis (40 patients)
 - Recovery : 67%
 - Average time for recovery : 3.3 \pm 2.8 days
 - 1 week of presentation in nearly all cases
 - Temporary pacing : 95% of cases
 - Permanent pacemakers in 27% (after 1 week)
 - Immunosuppression?

Batra et al. Pediatr Cardiol 2003;24:495-497

Rheumatic fever

- Asymptomatic rhythm and conduction abnormalities in children with acute RF
 - 64 children, 24 hr electrocardiography
 - First-degree AVB : 21.9%
 - Not related to the presence of carditis
 - Mobitz type I AVB : 1(1.56%)
 - Rheumatic affection of atrioventricular conduction is proximal to the trifascicular system ? Karacan et al. Cardiology in the Young 2010;20:620-630

Kawasaki disease

- T wave change, prolonged PR, QT interval, complete heart block
- Most prevalent in the first month of disease
- Do not predict the type of echocardiographic abnormalities



Duchenne's muscular dystrophy

- High incidence of ECG abnormalities in young pts with Duchenne's MD
 - 69 pts, aged \leq 18 yrs
 - Deep Q waves, low RV5 + SV1(91.3% of pts)
 - Initial and primary sites of myocardial dystrophy : posterobasal and contiguous left ventricular wall as
 - Dystrophin gene deficiency Takami et al. Pediatr Neurol 2008;39:399-403

Arrhythmia & obstructive sleep



Arrhythmia & obstructive sleep apnea

- a/w the number of apneic episodes and the severity of hypoxemia
- Nocturnal arrhythmia : 50% of pts
- Nonsustained VT, sinus arrest, second-degree AVB, PVC
- Prolonged apnea and hypoxemia → diving reflex → cardiac vagal activation → bradycardia, AVB and

Arrhythmia & obstructive sleep

apnea TABLE II

Cardiac Arrhythmia or Conduction Abnormality in 50 Patients With Sleep Apnea Syndrome Before and After Tracheostomy

	Before Tracheostomy		After Tracheostomy	
Cardiac Arrhythmia or Conduction Abnormality	Awake	Asleep	Awake	Asleep
Sinus arrest 4 to 13 s Second-degree atrioventricular block	0	15	0	0
Mobitz type I	0	5		0
Mobitz type II	0	10	0	0
Ventricular tachycardia	0	8	0.55	0.5
Atrial flutter	. 0	2	0	0
Atrial fibrillation	0	8	0	0
Extreme sinus bradycardia Frequent premature	0	2	0	0
ventricular contractions	11	1	1	

- Cardiac conduction system involvement in sudden death of obese young people
 - 7 patients, 5 obese and 2 mild to moderately obese
 - 6~32 years of age
 - 3 patients : OSA
 - Pathology





Fig. 1. Patient 1. Focal accumulation of mononuclear cells in approaches to SA node. *AP*, Approaches to the SA node. *Arrows* point to accumulation of mononuclear cells. (Hematoxylin-eosin stain—; original magnification ×200.)

Fig. 6. Patient 3. Arteriolosclerosis of AV node with fatty infiltration in adjacent ventricular septum. N, AV node; C, central fibrous body; F, fat in central fibrous body and ventricular septum; V, ventricular septum. Arrow points to





LBB

Fig. 5. Patient 3. Remnant of SA node with fibrosis and fatty infiltration. SA, Sinoatrial node; F, fat; FI, fibrosis. (Hematoxylin-eosin stain; original magnification $\times 40$.)

Fig. 2. Patient 1. Right ventricular infundibular muscle pressing on branching bundle, with focal fibrosis of branching bundle, mid part of left bundle branch, and ventricular septum. I, Infundibular septal bulge pressing on the branching bundle; B, branching bundle; LBB, mid part of left bundle branch. Arrows point to fibrosis. Weigert-van Gieson stain; original magnification $\times 24$.)



- Pathologic findings of conduction system were more marked in patients with obesity of longstanding duration and a history of obstructive sleep apnea
- May produce arrhythmias that may be silent in nature but may form a mileu for an arrhythmic event that end fatality during an altered Bharati et al. Am Heart J 1995;129:273-81 physiologic state

Anorexia nervosa

- Bradycardia, hypotension, prolonged QTc
- Kanbur, et al. 2009
 - 12 yr old girl
 - Second-degree AV block (Mobitz type I)
 - Intrinsic or complication of AN ?



QTc

- Predisposing factors to QT prolongation
 - Age, female gender, LVH, heart failure, myocardial ischemia, hypertension, DM, increased thyroid hormone concentraction, elevated serum cholesterol, high BMI, slow HR, electrolyte abnormalities (hypokalemia, hypomagnesemia), drugs



Long QT syndrome medications

- Class IA and III antiarrhythmics
- Antibiotics (macrolides and quinolones)
- Antidepressants (tricyclics and selective serotonin reuptake inhibitors)
- Antipsychotics (haloperidol and phenothiazines)
- Antiemetics(ondansetron and prochlorperazine)

LQTS – medication

Mechanism

- Inhibition of the KCNH2encoded HERG potassium channel
 prolongation of the action potential duration and a prolonged QT interval
- 10% : quiescent LQTSsusceptibility mutations



 Risk factors : advanced age, female gender, hypokalemia, bradycardia,

hypomagnesamiac(Bayl Univ Med Cent) 2010;23(3):250-255