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Clinical follow - up of patients.

Konrad Brockmeier

Complete AV Block from Fetus to Adolescent

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Nothing to disclose

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The clinician is trying to prevent any damage to the patient

...once AVB is diagnosed.

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Knowledge of the natural history of severe brady-arrhythmias comes from old studies performed before or at the beginning of the PM era.

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Left untreated,
congenital AV block is associated with a fetal and neonatal mortality between 14 and 34 %
(outcome of n=55; 1979 - 1989)

Schmidt KG et al. J Am Coll Cardiol 1991;17:1360–6

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Left untreated, fetal or neonatal death correlated significantly with the presence of structural heart defects

Prognosis very poor:

only 4 (14%) of 29 survived the neonatal period.

Schmidt KG et al. J Am Coll Cardiol 1991;17:1360–6

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AVB with structural heart defects

- left atrial isomerism
- atrio-ventricular discordance

Schmidt KG et al. J Am Coll Cardiol 1991;17:1360–6

Taketazu M et al. Am J Cardiol 2006;97: 720 –724

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The estimated overall mortality in i-AVB **without** pacing is estimated to be around 8–16 % in infants and half as much in children and adults

Buyon JP et al. J Am Coll Cardiol (1998) 31:1658–66

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Clinical characteristics and outcome of fetal AVB are generally evaluated separately in

- CHD-AVB
- i-AVB

Death in patients with untreated AVB is due to

- heart failure (HF) secondary to low cardiac output
- sudden cardiac death caused by prolonged asystole
- bradycardia-triggered ventricular tachy-arrhythmia.

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Without pacing support, it appears that the slow heart rates and associated higher stroke volumes probably drive the risk

Michaëlsson M, Jonzon A, Riesenfeld T (1995)
Isolated congenital complete atrioventricular block in adult life. A prospective study.

Circulation 92:442–9

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Diagnostic steps

- History
- Examination
- ECG
- Holter
- ECHO

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Diagnostic steps

- ventricular and atrial heart rate at presentation
- minimum ventricular heart rate
- arrhythmias
- pauses
- diameter of the cavities of the heart
- function

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Indications for PM placement are as follows:
for symptomatic children

- signs of heart failure
- left ventricular dysfunction
- wide QRS interval
- prolonged QT interval

Indications for PM placement are as follows:
for symptomatic children

CHD-AVB: ventricular HR <70 bpm (Class I, level of evidence B)

Indications for PM placement are as follows:

for asymptomatic neonates and infants, prophylactic pacing was indicated when the

- ventricular rhythm was <55 bpm

in asymptomatic children beyond 1 year of age

- average heart rate <50 bpm
- long pauses on 24 h recordings
- and/or frequent PVB

Villain E. Pacing Clin Electrophysiol 2008;31(Suppl 1):S17–20.
Paul T German guidelines on bradyarrhythmias 2010

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Diagnostic steps

steps repeated as appropriate

Large series of congenital AVB, reported 95% of second degree **block progressing** to complete block during fetal or postnatal life

Lopes LM et al. Circulation 2008;118:1268–75.

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Inherited progressive cardiac conduction disease (PCCD)

- progressive conduction abnormality
- with an otherwise structurally normal heart
- hereditary

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PCCD progressive conduction abnormality
linked to genetic variants in the ion channel genes
SCN5A, SCN1B, SCN10A, TRPM4, KCNK17
& genes coding for cardiac connexin proteins

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Holt-Oram syndrome (Tbx5 mutations)

- autosomal-dominant inherited disease
- radial ray upper limb abnormalities,
- cardiac septation defects,
- ...and cardiac conduction disorders

(may occur even in the absence of overt structural heart disease)

Baban A et al Am J Med Genet A 2014;164A:1419–24

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AVB and

- skeletal myopathies
- muscular dystrophies

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Lyme disease (borrelia infection)
complete heart block is usually reversible
with appropriate antibiotics

Forrester JD et al. Clin Infect Dis 2014;59:996–1000

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post-op AVB:

wait 7 - 10 days

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Without pacing support, it appears that the slow heart rates and associated higher stroke volumes

probably drive the risk

—-> implant of PM is life saving and decreasing morbidity

Michaëlsson M, Jonzon A, Riesenfeld T (1995)
Isolated congenital complete atrioventricular block in adult life. A prospective study.

Circulation 92:442–9