

A case of episodic congenital complete atrioventricular block

G N Lucas¹, M A L Faiz²

Sri Lanka Journal of Child Health, 2002; **31**: 25-6

(Key words: congenital complete atrioventricular block, CCAVB)

Introduction

Congenital complete atrioventricular block (CCAVB) occurs in 2 settings characterised by the presence or absence of major congenital anatomic cardiac defects¹. Certain forms of congenital heart disease have a high association with CCAVB, particularly AV septal defects and those involving abnormalities in the bulboventricular looping such as l-transposition of the great arteries. In this setting, heart block may occur either as the result of fibrous disruption between the atrium and the AV node or due to absence of the penetrating bundles of the AV node^{2,3}. CCAVB in the absence of structural heart disease occurs in 1/15,000-1/20,000 live births⁴. An association between maternal connective tissue disease and CCAVB was first demonstrated in 1977⁵.

Case report

A 4 year old boy from Panadura was admitted to the Lady Ridgeway Hospital in September, 2000 with repeated episodes of loss of consciousness from the age of 1 year, each episode lasting a few minutes and being accompanied by pallor and brief tonic-clonic seizure activity. Initially, epilepsy had been diagnosed and the child was treated with carbamazepine with little response. For the last 8 months he was not on anticonvulsant therapy.

On examination, the pulse rate was 72/min, regular and of good volume. The blood pressure was 100/60 mm Hg. There was a grade 2 ejection systolic murmur best heard in the pulmonary area with fixed splitting of the second heart sound. The chest x-ray showed pulmonary plethora and the ECG showed a partial right bundle branch block. A 2 D echocardiogram confirmed the diagnosis of secundum atrial septal defect.

Although the ECG did not show any evidence of complete heart block, because of the possibility that the seizures were really cardiac syncopal episodes (Stokes Adams attacks) the child was referred to the

cardiology unit for Holter monitoring. The Holter monitoring showed periods of complete block followed by 2:1 block (Figures 1 & 2). The child was transferred to the cardiology unit of the National Hospital of Sri Lanka for permanent cardiac pacing.

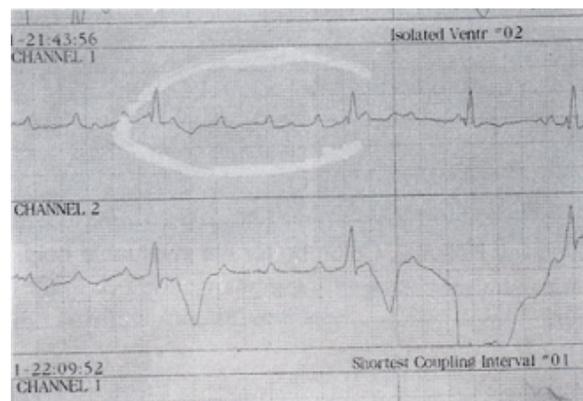


Figure 1 Holter monitoring showing periods of complete heart block

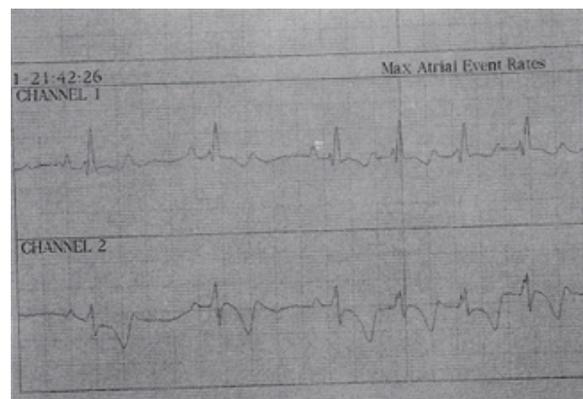


Figure 2 Holter monitoring showing 2:1 heart block

Discussion

Children with congenital complete atrioventricular block (CCAVB) may present at any age because of symptoms of bradycardia (fatigue or syncope). Many of these children are asymptomatic and are diagnosed because of a low resting heart rate on physical examination⁶. Our case is unusual in that the resting pulse rate was within normal limits. A surface ECG is usually all that is required to make the diagnosis of

¹Consultant Paediatrician, ²Paediatric Registrar, Lady Ridgeway Hospital, Colombo

CCAVB. In our patient the routine ECG did not show evidence of CCAVB and a 24-hour rhythm recording (Holter monitor) was necessary to establish the diagnosis. Cardiac syncopal episodes in CCAVB can be accompanied by brief tonic or clonic seizure activity which occur 10-20 seconds after the onset of asystole, is usually of short duration with no subsequent post-ictal phase⁷. It is not surprising that an initial diagnosis of epilepsy was made in our patient. The mainstay of treatment for symptomatic CCAVB is permanent cardiac pacing⁶.

Acknowledgements

We thank Dr S Narenthiran, cardiologist, Lady Ridgeway Hospital, Colombo for his invaluable help

References

1. Lev M. Pathogenesis of congenital atrioventricular block. *Prog Cardiovasc Dis* 1972; **15**: 145-57.
2. Lev M, Silverman J, Fitzmaurice F M, Paul M H, Cassels D E, Miller R A. Lack of connection between the atria and the more peripheral conduction system in congenital atrioventricular block. *Am J Cardiol* 1971; **27**: 481-8.
3. Lev M, Cuadoros H, Paul M H. Interruption of the atrioventricular bundle with congenital atrioventricular block. *Circulation* 1971; **43**: 703-10.
4. Michaels M, Engle M A. Congenital complete heart block: an international study of the natural history. *Cardiovasc Clin* 1972; **4**: 85-101.
5. Chameides L, Trex R C, Vetter V, Rashkind W J, Galioto F M Jr, Noonan J A. Association of maternal systemic lupus erythematosus with congenital complete heart block. *N Engl J Med* 1977; **297**: 1204-7.
6. Case C L. Diagnosis and treatment of pediatric arrhythmias. *Pediatr Clin North Am* 1999; **46**: 347-54.
7. Lewis D A, Dhala A. Syncope in the pediatric patient - the cardiologist's perspective. *Pediatr Clin North Am* 1999; **46**: 205-19.

