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Brief Report

Atrioventricular block in a newborn with acquired long QT syndrome

John R. Phillips, Christopher L. Case, Paul C. Gillette

Division of Pediatric Cardiology, Cook Children's Heart Center, Fort Worth, TX, USA

Abstract We report a case of 2 : 1 atrioventricular block associated with acquired long QT syndrome. A newborn presented to our neonatal intensive care unit with intermittent bradycardia due to 2 : 1 atrioventricular block. Initial evaluation showed QT prolongation and significant electrolytic abnormalities. After correction of the electrolytic imbalance, the QT interval normalized and atrioventricular block resolved. Compared to congenital long QT syndrome with 2 : 1 atrioventricular block, acquired long QT syndrome with comparable atrioventricular block has a benign prognosis, provided treatment is initiated quickly.

Keywords: Acquired long QT syndrome; atrioventricular block

THE LONG QT SYNDROME IS CHARACTERIZED BY prolongation of the QT interval and ventricular arrhythmias. In its more dramatic form, P waves may fall within the previous T wave and fail to conduct to the refractory ventricles. In these cases, there is 2 : 1 atrioventricular block. The combination of congenital long QT syndrome and 2 : 1 atrioventricular block in childhood portends a poor outcome, with half of reported patients dying in infancy and early childhood.[1,2] Less is known about the prognosis of infants with 2 : 1 atrioventricular block secondary to acquired long QT syndrome. We report such a case of acquired long QT with 2 : 1 atrioventricular block secondary to significant electrolytic abnormalities. QT prolongation and atrioventricular block resolved with correction of the electrolytic imbalance. The prognosis of infants with acquired long QT and 2 : 1 atrioventricular block is benign compared to infants with the congenital syndrome.

Case report

A female, born at 36 weeks gestation of an insulin-dependent diabetic mother, was transferred to our institution because of tachypnea, metabolic acidosis, and abrupt episodes of bradycardia. She had been treated with Solu-Medrol, hydrocortisone and Diazoxide prior to transfer. At presentation, she had mild respiratory distress with a metabolic acidosis. Telemetric monitoring showed normal sinus rhythm with intermittent 2 : 1 atrioventricular block. There was no family history of congenital heart disease, long QT syndrome, connective tissue disorders, or sudden death.

Her physical exam was unremarkable. In general, she was acyanotic with mild respiratory distress. The lungs were clear to auscultation bilaterally. She displayed abdominal breathing without intercostal retractions or grunting. Chest expansion was symmetrical. Auscultation of the heart revealed normal sounds with physiologic splitting. There were no murmurs, rubs or gallops. The abdomen was soft without hepatosplenomegaly. Pulses were brisk and equal in all limbs. There was no peripheral clubbing or edema.

Her initial electrocardiogram showed normal sinus rhythm with 2 : 1 atrioventricular block (Fig. 1) and occasional monomorphic premature ventricular contractions. The corrected QT interval measured 533 ms using Bazett's formula ($QTc = QT\sqrt{RR}$).[3] The S-T segments were long with late-onset T waves. There was evidence of biventricular hypertrophy. Her initial echocardiogram revealed normal

Correspondence to: John R Phillips, MD, Pediatric Cardiology, Robert C Byrd Health Sciences Center, PO Box 9214, Morgantown, WV 26506-9214, USA. Tel: 304 293 7036; Fax: 304 293 1409; E-mail: jphillips@hsc.wvu.edu

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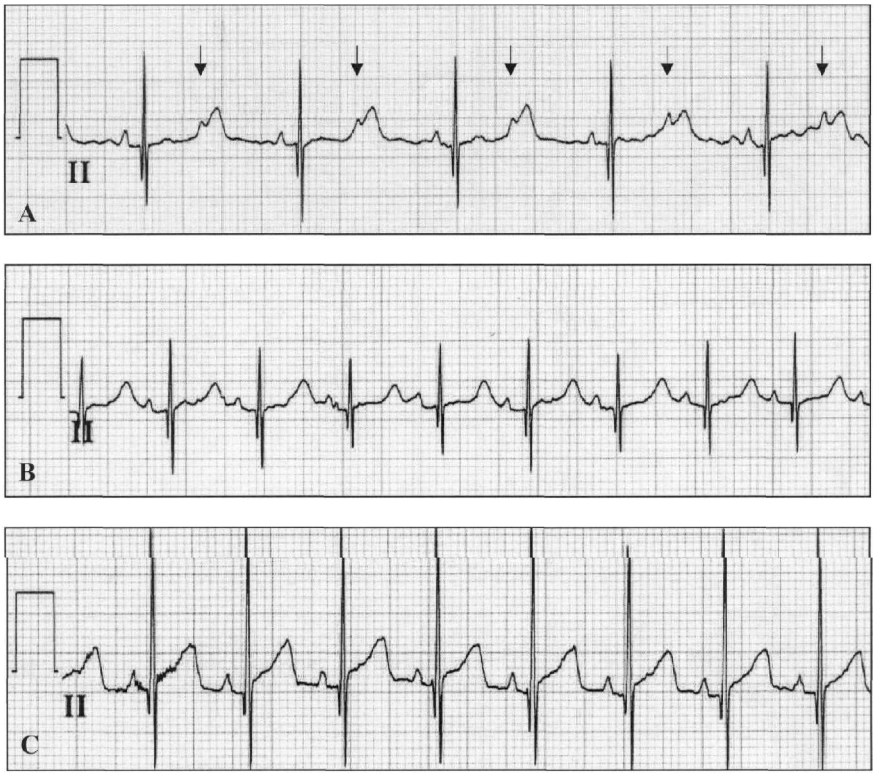

Figure 1.
Progression of electrocardiograms in our patient; paper speed, 25 mm/sec. (A) Presenting electrocardiogram: sinus rhythm with 2:1 atrioventricular block, QTc = 533 ms. Arrows indicate blocked P waves. (B) Electrocardiogram 2 days later: sinus rhythm, QTc = 489 ms. (C) Electrocardio- gram 7 weeks later: sinus rhythm, QTc = 410 ms.

anatomy, with concentric biventricular hypertrophy and hyperdynamic wall motion. There was no obstruction to the ventricular outflow tract. Left-to-right shunting was seen across a patent oval foramen. Blood analysis demonstrated several electrolytic abnormalities, including hypocalcemia, hypokalemia, and hypernatremia.

Acute treatment consisted of supplemental calcium and sodium bicarbonate and discontinuation of her prior medications. Over the next 2 days, her electrolytes normalized and acidosis corrected. Simultaneously, her conduction defect resolved and the corrected QT interval shortened to 489 ms (Fig. 1). She was monitored extensively during her admission and had no ventricular arrhythmias more complex than single monomorphic premature ventricular contractions. She was prophylactically treated with propranolol (3 mg/kg/day). She remained hemodynamically stable throughout her hospitalization and was discharged to home. Follow-up evaluation at 7 weeks of life was unremarkable. She was feeding well with appropriate growth. Her rhythm was normal, as was her corrected QT interval at 410 ms (Fig. 1). Holter monitoring showed normal sinus rhythm with rare monomorphic premature ventricular contractions. Repeat echocardiography showed resolution of her ventricular hypertrophy.

Discussion

In addition to being congenitally transmitted, the long QT syndrome can be acquired. Factors associated with the acquired form include metabolic abnormalities such as hypokalemia and hypomagnesemia, starvation as in anorexia nervosa, cardiac ischemia, and medications such as general anesthetics, antihistamines, and antiarrhythmics. In our case, we believe that the acquired syndrome was due to significant electrolytic disturbances. Interestingly, her abnormality led to 2:1 atrioventricular block, a finding seldom reported with the acquired syndrome. In reviewing the literature, we found only one other reported case of acquired long QT syndrome with 2:1 atrioventricular block.[4] In that instance, QT prolongation was a result of cisapride usage and resolved with discontinuation of the drug.

It has been shown that 6% of patients with the long QT syndrome have a normal corrected QT interval.[5] We wonder if these patients have subclinical abnormalities of the ion channels that are unmasked by electrolytic imbalance or drugs. Of this we are uncertain. Erring on the side of caution, nonetheless, we have recommended that our patient avoid drugs which might prolong the QT interval, and have serial electrocardiograms to monitor this

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interval. Further, at the time of presentation, we chose prophylactically to treat with propranolol in light of her abnormality, concentric ventricular hypertrophy, and premature ventricular contractions. Ventricular hypertrophy is commonly seen in infants of diabetic mothers, however, so congenital hypertrophic cardiomyopathy could not be ruled out in our patient. We will likely discontinue medication at 1 year of age if she remains normal.

In conclusion, previous studies have shown that the congenital long QT syndrome with 2:1 atrioventricular block in infants and children is often a fatal combination. Our case suggests that an acquired long QT interval with 2:1 atrioventricular block has a benign prognosis, provided therapy to reverse the prolongation of the QT interval is initiated quickly. We suggest long-term follow-up of these patients to identify covert cases of the congenital syndrome.

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