Original Articles

Incidence and Significance of Primary Abnormalities of Cardiac Rhythm in Infants at High Risk for Sudden Infant Death Syndrome

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SUMMARY. The exact relationship between cardiac arrhythmias and sudden infant death syndrome (SIDS) is uncertain. Several reports have implicated both ventricular and supraventricular arrhythmias in isolated cases, but there have been no studies of the incidence or type of arrhythmias that occur in populations at risk for SIDS. Of 1699 infants at high risk for SIDS, 60 (4%) were found to have a primary cardiac arrhythmia (i.e., not associated with disordered respiration or apnea). The incidence of atrial and ventricular premature beats, supraventricular tachycardia, and Wolff-Parkinson-White syndrome was similar to the incidence found in normal infants. Primary bradycardia (defined as a heart rate less than 60 for greater than 10 s not associated with abnormal respiration) was the most common arrhythmia, occurring with a frequency and severity not seen in normal infants. Thirty-two infants experienced periodic bradycardia. In 19 of these latter infants, there were symptoms associated with these bradyarrhythmias that necessitated treatment. Heart rates as low as 20 beats/min were recorded. One infant presented with an episode of ventricular fibrillation and on further evaluation was noted to have recurrent bradyarrhythmias. In no infant was there abnormal prolongation of the QT interval.

Primary bradyarrhythmias are seen at an increased incidence in infants at high risk for SIDS and may play a causal role in this syndrome. Most symptomatic infants can be adequately controlled with sympathomimetic or parasympatholytic therapy. Other cardiac arrhythmias occur at a rate similar to that in normal infants and are therefore unlikely to play a major role in SIDS.

KEY WORDS: Cardiac arrhythmias — Parasympatholytic therapy — Sudden infant death syndrome — sympathomimetic therapy

Sudden infant death syndrome (SIDS) is the sudden unexpected death of a previously well infant less than one year of age, for which there is no clinical or pathologic explanation in spite of an autopsy. This syndrome accounts for 8000 infant deaths annually in the United States, making it the leading cause of death between one month and one year of life [10, 25]. Although intensive study has delineated some of the demographic and pathologic characteristics of this syndrome [39], the precise cause (or causes) of SIDS is still unknown. Reasoning from the adult experience with sudden death, it has been postulated that at least some of these sudden infant deaths are due to ventricular fibrillation [6].

To examine this hypothesis, we studied a population of infants who have been shown to have a high risk for SIDS (HRSIDS) including: (a) “near SIDS” infants (infants who are found apneic, cyanotic, limp, and unresponsive during sleep and are successfully resuscitated), (b) siblings of SIDS infants, and (c) preterm infants.

In this study, we report our findings of the incidence, type, significance, management, and follow-up of primary cardiac arrhythmia in the popula-
tion defined as being at high risk for SIDS. We also review and discuss the literature pertinent to this problem.

Materials and Methods

Between 1975 and 1979, investigations were performed on 1699 infants referred as HRSIDS infants for evaluation at the Massachusetts General Hospital. All patients were less than one year of age at the time of initial evaluation. Infants defined as HRSIDS were evaluated on the basis of one or more of the following categories: (a) near SIDS who have an estimated 10% incidence of SIDS [15], (b) siblings of a previous victim of SIDS among whom a 2% incidence of SIDS has been reported [39], and (c) premature infants who have a 2%-2.5% incidence of SIDS [23]. One or more 12-h continuous recordings of cardiac rhythm and respiratory activity by impedance pneumography were obtained on each subject. Patients found to have abnormalities of cardiac rhythm not associated with altered respiratory patterns (i.e., prolonged apnea or disorganized, shallow, or periodic breathing) were evaluated to exclude underlying structural heart disease. In these infants, evaluation included pre- and postnatal histories, general and cardiac physical examination, standard 12-lead electrocardiograms and, when appropriate due to other coexisting problems or uncertainty concerning the absence of structural heart disease, posterior–anterior and lateral chest roentgenograms, serum electrolytes and calcium determinations, arterial blood gases, cardiac ultrasound, and continuous 24-h cardiac monitoring. It should be noted that the normal heart rate in infancy varies considerably with activity, but is usually between 110 and 140 beats/min. Bradycardia in infancy is usually defined as heart rates less than 90 [2, 7, 42]. To exclude sinus arrhythmia and transient sinus node slowing, which occur frequently in this patient population, we defined bradyarrhythmia as a heart rate less than 60/min persisting for 10 s or longer. Corrected QT interval was determined as previously described [15] by dividing the QT by the square root of the RR interval, determined during periods of normal sinus rhythm. Atrial and ventricular premature beats, supraventricular tachycardia, heart block, and ventricular fibrillation were defined by standard criteria [7].

Duration of follow-up varied from one to 72 months. The study end-point was defined by resolution of the arrhythmia on subsequent clinical and long-term cardiac rhythm monitoring or, when persistence of arrhythmia was noted, the follow-up interval was continued to the present time.

Results

A total of 60 infants, or 4% of the 1699 "high risk" infants, were recognized to have cardiac arrhythmias in the absence of structural heart disease or disordered respiratory pattern. Table 1 lists the type and incidence of primary cardiac arrhythmias observed in our study population. No cases of long QT interval syndrome were identified.

Bradyarrhythmias were most common in the preterm infant group. Ten of these latter infants who experienced bradyarrhythmias had a gestational age of 26–32 weeks, six of 32–35 weeks, four of 36–38 weeks, and 12 infants were full term. This was the only arrhythmia for which prematurity was a risk factor. In addition to persistent heart rates below 60 in all 32 patients, rates less than 50 were seen in 18 patients and rates as low as 20 beats/min occurred in two infants with spontaneous return to a normal heart rate. The 14 patients with minimum rates of 50–60 had junctional escape rhythms during periods of sinus bradycardia, whereas the patients with bradycardia below 50 either remained in sinus rhythm with no junctional escape rhythm or manifested slow junctional rhythm only after sinus rate fell below 30–40 beats/min, suggesting a slow junctional escape mechanism. In all, there was a normal QRS interval. A total of 19 infants experienced recurrent bradyarrhythmia associated with signs of hypoperfusion and cyanosis, and were treated. Of these, 15 received sympathetic agonists (theophylline in 12 and caffeine in three) and four were treated with parasympathetic antagonists (atropine sulfate in three and belladonna in one). Bradycardia resolved in 12 during treatment and persisted with improvement in seven. After 3–36 months of follow-up, 25 infants were arrhythmia free and seven infants with persistent bradyarrhythmias are still followed. Three of these latter infants were preterm, one was a sibling of an SIDS victim, and three were initially evaluated as near SIDS (with bradyarrhythmias first detected at eight weeks, nine weeks, and 13 months of age). Of note, in a cohort of 131 normal infants investigated by the same methods, in no instance did we record a heart rate below 60/min.

Atrial premature beats (APBs) were seen in 16 infants. In eight infants, 10 APBs/min were found, in seven infants, 10–20 APBs/min occurred, and one infant was noted to have 20 APBs/min during sleep. After 3–20 months of follow-up, four infants continue to experience APBs, whereas 12 infants returned to normal rhythm within 1–6 months. No patients with APBs had symptoms or related complications and none were treated.

Ventricular premature beats (VPBs) were found in six infants, one of whom continues to have ectopic beats after six months of follow-up. Again,