The short QT syndrome and sudden infant death syndrome

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A three-week-old neonate with a shortened QT interval of 210 ms was admitted to the Southwest SIDS Research Institute (Texas, USA) for monitoring because of apnea. A wandering (shifting) pacemaker was present on admission (Figure 1), which was recognized as a frequent antecedent to symptomatic sinus node dysfunction (SND). There was also ST segment elevation and negative U waves (1). Finally, a 2:1 sinoatrial exit block was documented along with a junctional escape beat subsequent to vagotonic sinus arrest (2). With progressive default of the primary pacemaker the escape rhythm, atrioventricular dissociation developed (Figure 2). Asystole ensued following the development of critical SND: irregular sinus bradycardia consequent on episodic sinus inertia and/or third-degree sinoatrial exit block (Figure 3).

Recently, the short QT syndrome (SQTS), a cardiac ion channelopathy has been cited in the pathophysiology of cardiac arrest in sudden infant death syndrome due to idiopathic ventricular fibrillation (IVF) (3). Further studies are needed to delineate the exact incidence and role of SQTS in relation

to IVF (4). In the present case, sudden infant death syndrome with SQTS was recorded, in which SND was an important aspect of the pathophysiology.

In summary, a postdelivery electrocardiogram with an abbreviated QT interval has both clinical and genetic (5) implications in this primary electrical disorder in that it may be the harbinger not only of IVF, but also of disordered cardiac impulse formation in a potentially sick sinus node.

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Figure 1) Electrocardiogram lead 2 of a patient with short QT syndrome. Note ST-segment elevation, negative U waves and peaked T waves. Figure courtesy of Dr Richard Hardoin at the Southwest SIDS Research Institute, Texas, USA

Figure 2) Atrioventricular dissociation – 'marching through' phenomenon. Figure courtesy of Dr Richard Hardoin at the Southwest SIDS Research Institute, Texas, USA

Figure 3) Terminal event. Episodic atrial and ventricular standstill with sinus inertia and/or third-degree sinoatrial exit block. Figure courtesy of Dr Richard Hardoin at the Southwest SIDS Research Institute, Texas, USA

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