

Life threatening QT prolongation associated with congenital hydrocephalus in a preterm infant.

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INTRODUCTION: Temporary QT-interval prolongation following intracranial haemorrhage and hydrocephalus has been repeatedly reported in adults. CASE: We present the first case of a preterm infant of 31 weeks gestational age suffering from intrauterine aqueductal stenosis. Postnatal cardiopulmonary adaption was unremarkable. Serum electrolytes were within normal limits and a cranial ultrasound showed hydrocephalus without signs of elevated intracranial pressure. On day two of life he developed sudden bradycardia with poor peripheral perfusion and mild arterial hypotension, which resolved spontaneously after 10 minutes. ECG showed a sinus rhythm with 70 bpm and an excessive QT prolongation (QTc 560ms) resulting in 2:1 atrioventricular conduction due to ventricular refractoriness as reported in LQT patients. Noticeably, the AV nodal tissue is unaffected and AV conduction is normal in this patient. Several episodes of bradycardia occurred until day 5 of life when they finally stopped. Family history of LQT syndrome was negative and the identical twin brother had repetitive normal ECG`s. DISCUSSION: This case illustrates a potentially life threatening QT prolongation associated with congenital hydrocephalus. The cause of the temporary QT prolongation remains unclear. Sympathetic-parasympathetic imbalance or electrolyte fluctuations are reported theories in adults. As this association has never been reported in a neonate before, it remains notional if the QT-interval was influenced rather by an intermittent intracranial pressure elevation, or if there was a hydrocephalus caused impairment of the sympathetic-parasympathetic balance during the process of perinatal adaption. The authors encourage hemodynamic monitoring of these patients as the reported phenomenon occurred suddenly, no data exists to identify affected individuals and serious hemodynamic compromise has to be expected in case of a longer-lasting episode. CONCLUSION: This is the first report of excessive, life threatening QT prolongation associated with congenital hydrocephalus in a preterm infant.