Introduction Alternating hemiplegia of childhood (AHC) is a rare neurodevelopmental disorder caused by de novo mutations in the ATP1A3 gene encoding the $\alpha 3$ -subunit of the Na⁺/K⁺-ATPase transporter, expressed in cardiac and neuronal tissue. AHC is characterised by recurrent plegic attacks, dystonic posturing and seizures. Non-paroxysmal neurological manifestations and developmental delay may occur. Potentially fatal arrhythmias are associated with cardiac channelopathies but also with neurological channelopathies such as Andersen-Tawil syndrome, and postulated in sudden unexplained death in epilepsy. Autonomic dysfunction and one case of asystole have been reported in AHC, but specific cardiac abnormalities have not been defined.

Methods and results Nine patients from an original UK cohort of twenty-six (adults n=6; children n=3), underwent detailed cardiac and neurological phenotyping, baseline ECG testing and ATP1A3 molecular genetic analysis. Repolarisation abnormalities and varying degrees of intraventricular conduction delay were seen in all six, ATP1A3-mutation bearing, adults. No abnormalities were detected in children.

Conclusions These cardiac features raise the possibility of an arrhythmogenic potential of a neuronal channelopathy co-expressed in the heart and brain, warranting cardiac surveillance from adolescence into adulthood and further risk prediction strategies in AHC.

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CARDIAC FEATURES IN ADULTS WITH ALTERNATING HEMIPLEGIA

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