Children's ajmaline test

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Ajmaline testing appears safe and feasible in children when performed in an appropriate setting by an experienced team.

Test positivity may change with age in individuals, suggesting that the test should be repeated in the late teenage years or early adulthood. (**Merlin Ranald McMillan, Thomas George Day, Margarita Bartsota, Sarah Mead-Regan, Rory Bryant, Jasveer Mangat, Dominic Abrams, Martin Lowe, Juan Pablo Kaski Feasibility and outcomes of ajmaline provocation testing for Brugada syndrome in children in a specialist paediatric inherited cardiovascular diseases centre Open Heart. 2014 Feb 12;1(1):e000023. doi: 10.1136/openhrt-2013-000023. eCollection 2014**.)

Prevalence of BrS in the paediatric population is extremely low (0.009%) as compared with the adult population (0.14–0.7%). (**Yamakawa Y, Ishikawa T, Uchino K, et al. Prevalence of right bundle-branch block and right precordial ST-segment elevation (Brugada-type electrocardiogram) in Japanese children. *Circ J* 2004;68:275–9**) (**Hermida JS, Lemoine JL, Aoun FB, et al. Prevalence of the Brugada syndrome in an apparently healthy population. *Am J Cardiol* 2000; 86:91–4**)

Moreover, the mean age of patients presenting with either symptomatic or asymptomatic BrS has been reported to be in the fourth or fifth decade.

(**Priori SG, Napolitano C, Gasparini M, et al. Natural History of Brugada Syndrome: insights for Risk Stratification and Management. *Circulation* 2002;105:1342–7**)

Although rarely diagnosed in children, BrS can manifest at every age and may cause sudden death in childhood even in the first month of life. (**Priori SG, Napolitano C, Giordano U, et al. Brugada syndrome and sudden cardiac death in children. *Lancet* 2000; 355:808–9**)

Therefore, patients with suspected BrS and non-diagnostic baseline ECG should undergo ajmaline test clinical circumstances and wishes of family.

Performing ajmaline challenge for suspected BrS has been considered to be a safe procedure if drug discontinuation criteria are followed. (**Rolf S, Bruns HJ, Wichter T, et al. The ajmaline challenge in Brugada syndrome: diagnostic impact, safety, and recommended protocol. *EHJ* 2003;24:1104–12**).

However, specific data on the effects and safety of ajmaline test in individuals younger than 12 years are lacking.

Recently, Conte, Sieira and Pedro Brugada reported an incidence of ajmaline-induced sVA in 4.4% of patients younger than 18 years with ajmaline-induced BrS (**Conte G, Sieira J, Sarkozy A, et al. Life-threatening ventricular arrhythmias during ajmaline challenge in patients with Brugada syndrome: incidence, clinical features, and prognosis. *Heart Rhythm* 2013;10:1869–74**)

Experiencing an arrhythmic event during a diagnostic drug challenge could have tragic consequences, particularly if the patient is a child with BrS.

Although the occurrence of episodes of sVA might be significantly lower in a subject without a sodium channelopathy, **ajmaline challenge should always be performed under close supervision in an appropriate environment with all advanced life support facilities available, ideally including the possibility of performing a venoarterial extracorporeal membrane oxygenation placement in case of an intractable episode of ventricular fibrillation. Further studies conducted in more selected patients' populations (<12 years) could be helpful to clarify these important aspects, concerning the safety of ajmaline challenge in children with suspected BS.**

Clinical aspects and prognosis of either spontaneous or drug-induced BrS have been previously described in individuals younger than 16 years, and, as in adults, a higher risk of arrhythmic events has been found in symptomatic patients and in those displaying a spontaneous type I ECG. (**Probst V, Denjoy I, Meregalli P, et al. Clinical aspects and prognosis of Brugada syndrome in children. *Circ* 2007;115:2042–8**)

No systematic studies have yet been carried out to assess the clinical characteristics and the long-term follow-up of drug-induced BrS in patients younger than 12 years.
Screening of the family of a proband with BrS is always recommended.
However, the ideal age for screening of BrS family members is unknown and different centres use different protocols for the screening of the pediatric first-degree relatives.
Moreover, the clinical significance of repeating ajmaline challenge after puberty in pediatric family members of BrS patients with an initial negative drug test is unknown.
Hormonal changes can play an important role in the clinical presentation of BrS.
In particular, testosterone has been suggested as a potential hormone responsible for the age-dependent manifestation of BS phenotype, as suggested by the disappearance of Brugada type I ECG after surgical castration for prostate cancer. (**Matsuo K, Akahoshi M, Seto S, et al. Disappearance of the Brugada-type electrocardiogram after surgical castration: a role for testosterone and an explanation for the male preponderance. *Pacing Clin Electrophysiol* 2003;26:1551–3**)
The existence of an age-dependent response to ajmaline challenge is intriguing and might have relevant clinical implications.

However, repeating ajmaline challenge after puberty in patients with an initial negative drug test remains controversial and should be further investigated. (**Giulio Conte, Pedro Brugada. The challenges of performing ajmaline challenge in children with suspected Brugada Open Heart. 2014 Feb 20;1(1):e000031. doi: 10.1136/openhrt-2013-000031. eCollection 2014. syndrome**)