Cardiology in the Young

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Deglutition syncope to gastronomic satiety: a paediatric case report and update on therapeutic options

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Case Report

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Abstract

We describe a rare and severe presentation of deglutition syncope, manifesting as complete heart block, along with newer therapeutic options, including cardioneural ablation and leadless pacing. Our 15-year-old patient presented with frequent syncope with swallowing, along with symptoms of orthostatic intolerance and anxiety. When standard non-pharmacologic and pharmacologic treatments were insufficient, cardioneural ablation resulted in improvement in syncope. Subsequent standard treatment of orthostatic intolerance has significantly improved the quality of life, including allowing her a more normal diet. The option of leadless pacing to prevent bradycardia during episodes of induced heart block has not yet been enacted due to her clinical improvement.

Introduction

In a world of confusion and strife, all can agree that macaroni and cheese should be a pasta dish to enjoy. For our patient, prior to presentation, however, it led not to satiety but instead to syncope. This case report describes the endeavours to set this right.

Case report

A 15-year-old female presented to the clinic with a myriad of symptoms, including consistent syncope with swallowing solid foods. The episodes had occurred for many years, but had escalated in frequency and intensity over the six months prior to presentation. Macaroni and cheese was particularly likely to precipitate an event. She additionally had symptoms of orthostatic intolerance, including positional dizziness, fatigue, palpitations, and headaches. Non-pharmacologic treatment of orthostatic intolerance was recommended, including optimising water and salt intake and treating contributing conditions—she was found to be both iron and vitamin D deficient and started on supplementation. Given her severe presentation of recurrent syncope, a Holter monitor was placed.

Over 24 hours, multiple episodes of syncope occurred with swallowing food. Figure 1 shows an electrocardiographic tracing during an event, specifically demonstrating complete atrioventricular heart block. Vagally mediated atrioventricular block was suspected, given the preceding prolongation of the PR interval prior to complete block.

Given this finding, aggressive medical therapy was implemented, including starting fludrocortisone for orthostatic intolerance and glycopyrrolate for its anticholinergic properties, in hopes of preventing vagally mediated heart block. At initial dosing (1 mg by mouth three times per day prior to meals), glycopyrrolate was ineffective, and with increasing doses (2 and 3 mg per dose), bothersome side effects, including dry mouth and constipation, precluded ongoing use.

Workup with gastroenterology consultation was completed to be sure no primary precipitating pathology was present. Contrast upper gastrointestinal fluoroscopy was normal, as were esophagogastroduodenoscopy and biopsy. Due to her recalcitrant symptoms, additional interventions were considered, including leadless pacemaker placement within the right ventricle (Medtronic Micra™) to circumvent episodes of heart block and electrophysiologic study and cardioneural ablation. After meeting with the electrophysiology team, the family elected for ablation.

During the procedure, three-dimensional electroanatomic mapping (Carto-3, Biosense-Webster, Diamond Bar, California) of the right and left atria was performed. High-frequency stimulation from the ablation catheter was performed at anatomic regions with a high likelihood of ganglionic plexi. Areas with a vagal response, defined by at least a 50% increase in the R-R interval after high-frequency stimulation, were marked in the mapping system. Following the diagnostic portion of the procedure, radiofrequency ablation was performed at regions deemed to be a positive vagal response. If there was a risk of phrenic nerve damage, high output pacing at the intended target site was performed prior to ablation to ensure safety. Ablation was

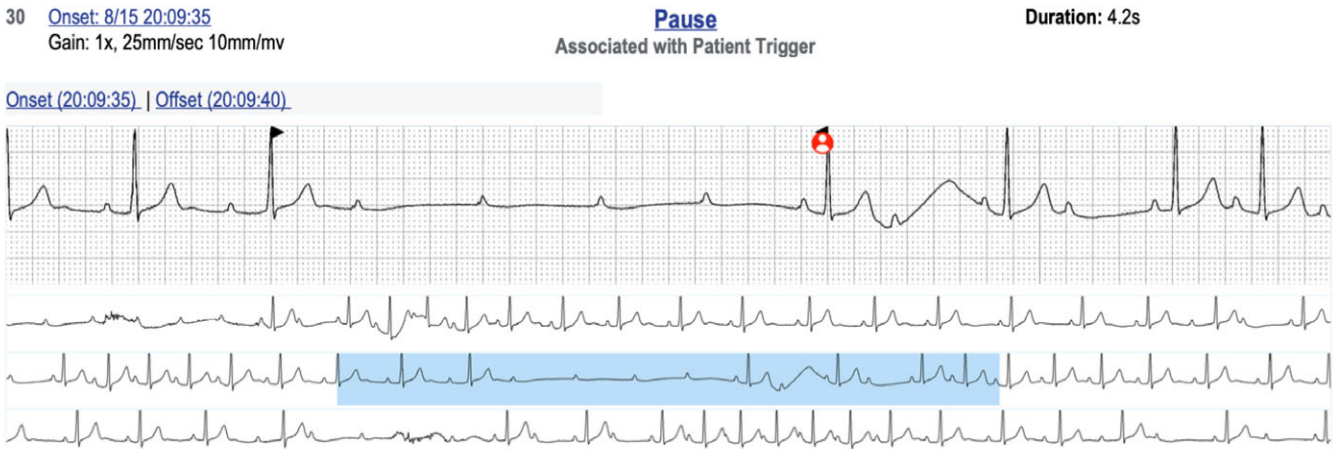

Figure 1. Holter monitor single lead tracing demonstrating complete heart block preceded by lengthening of the PR interval.

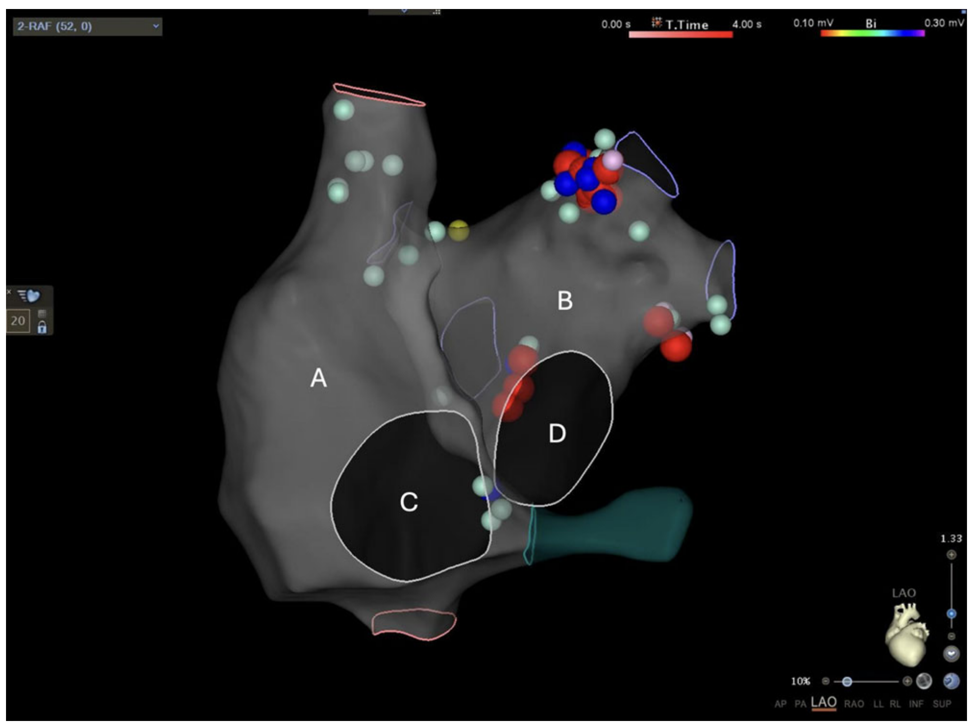

Figure 2. Electroanatomic map demonstrating the right atrium (***a***), left atrium (***b***), tricuspid valve (***c***), and mitral valve (***d***). Dark blue lesions indicate locations with an evoked vagal response. Light blue lesions indicate locations without an evoked vagal response. Red lesions indicate locations where RF ablation was performed.

performed at 45W through an open irrigated catheter with a target ablation index of 500 except where there was proximity to the oesophagus, where the index target was lowered to 380. In total, ganglionic plexi were targeted in proximity to the left superior, left inferior, and right inferior pulmonary veins (Figure 2). There was no positive response outside of the ostium of the right superior pulmonary vein, so this location was not targeted. There was no ablation in the right atrium. Following ablation, there was a decrease in sinus rhythm cycle length to 900 msec, and the PR interval was shortened, indicating tempering of intracardiac parasympathetic tone. Electrophysiologic measurements thereafter were:

AH = 112 ms

HV = 47 ms

RVP—Midline, concentric, decremental conduction with a VAWCL of 800 ms.

RAP—No evidence of PR > RR with an AVWCL of 1000 ms.

There were no complications with the procedure.

With follow-up, the family noticed improvement in syncope frequency, but other symptoms, including dizziness, fatigue, and headaches, remained bothersome. However, it became apparent that renewed focus on the medical orthostatic intolerance plan was needed, especially prior to considering pacemaker placement or a repeat attempt at ablation. With improved compliance with her plan, syncope resolved, with no episodes over the three months prior to her most recent clinic visit. She no longer has bothersome dizziness, and her headaches have resolved. As far as her diet, she has chosen to modify some food choices that were seemingly more

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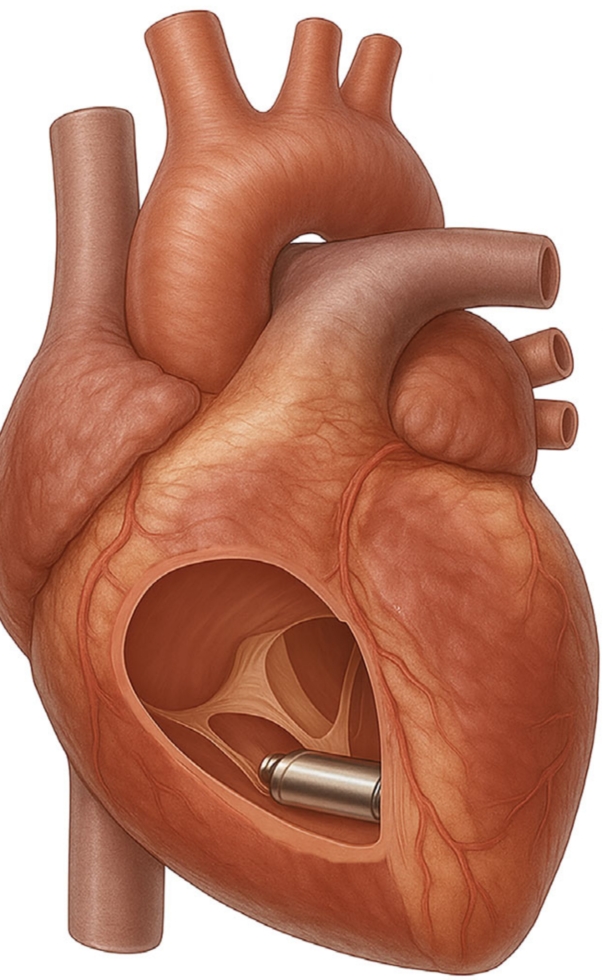

Figure 3. Leadless pacemaker device deployed within the right ventricle. Generated by Austin Kane utilising OpenAI, ChatGPT, October 2025.

likely to cause syncope. She avoids some harder-textured foods; however, she admits that when she forgets this plan, she can often have no symptoms with foods that triggered syncope in the past. On the other hand, she sometimes feels dizzy with softer-textured foods, such as a single pasta noodle. Anxiety about episode recurrence likely plays a role in this uneven presentation. Her self-assessed quality of life, as rated on the most recent clinic visit intake form, is now 10 out of 10 compared to 5 out of 10 at presentation. The family does not wish to move forward with a repeat EP study or pacing at this time. Perhaps most importantly, she has now been able to enjoy full servings of macaroni and cheese.

Discussion

The anatomy of the vagus nerves as they course through the thorax, as well as the cardiac autonomic nervous system anatomy, is germane to the pathophysiology and treatment described. Within the chest, the right vagus nerve runs posterior to the superior vena cava and right mainstem bronchus and then innervates cardiac and oesophageal plexuses, providing parasympathetic input to the heart and efferent (smooth muscle) and afferent (sensory) fibres to the oesophagus.[1] The left vagus nerve descends into the thorax between the left common carotid and subclavian arteries, gives rise to the left recurrent laryngeal nerve, and then provides innervation to both cardiac and oesophageal plexuses.[1] Thus, there is anatomic basis for parasympathetic reflex activation in response to strong afferent signalling from the oesophagus, i.e. significant visceral distension or an underlying pathologic process.[2] The intrinsic cardiac nervous system includes ganglionated plexi, which exert preferential control over specific areas within the heart.[3] The left atrial ganglia (near the pulmonary veins) primarily affect atrioventricular node function and thus are interventional targets in patients with inducible heart block.[4]

Although deglutition (swallow) syncope in adults was first described in the 18th century,[5] the first case report in paediatrics was not published until 1986.[6] The relative paucity of data in children likely has to do with increased symptom burden in adults, due in turn to a higher prevalence of contributing baseline cardiac and oesophageal disease. While a “cornucopia” of treatment options has been proposed for adults, including diet modification,[7] medications,[7] cardioneural ablation,[8] and pacing,[9] treatment proposals for children are sparse. In the most recent paediatric case report on the subject, the authors utilised a transvenous pacemaker[10] and commented on only 3 weeks of follow-up. Much has been learned from the adult experience since that time and deserves discussion.

Cardioneural ablation for parasympathetically induced bradyarrhythmias, including atrioventricular block, was first reported in 2005.[11] Over time, the importance of the posteromedial left ganglionic plexus in patients with AV block has been emphasised; while the superior right plexus principally modulates the sinus node, this leftward plexus in close anatomic proximity to the AV node is thought to be its final direct parasympathetic input.[4] Aksu et al. reported a case series of 31 patients with successful treatment of functional atrioventricular block via ablation targeting the posteromedial left ganglionic plexus.[12] Furthermore, results from a recently published multicenter trial, encompassing 15 centres and 205 adult patients (mean age 47 ± 17 years), are encouraging, with less than 5% minor complication rate, 78% of patients free of syncope at over 1 year of follow-up, and 97% freedom from pacemaker implantation.[13] By contrast, there are only a few reports of the use of cardioneural ablation in children, with a recent single-centre series describing only six patients with excellent results at 6-month follow-up.[14] Complications and long-term maladaptive effects are also a concern in paediatric patients, with recent reports of post-ablation inappropriate sinus tachycardia hopefully not foretelling of further complications from attenuated parasympathetic tone on the young heart.[15,16]

Compared to the “slim-pickings” of data on paediatric cardioneural ablation, a “smorgasbord” of reports on ventricular pacing for inducible atrioventricular block has been presented.[17–21] Even in clinical trials, however, pacing is often reported to incompletely treat syncope.[22] Hemodynamically this would be expected, as a vasodepressor component to symptoms (vasovagal syncope) often is present instead of pure cardioinhibition.[23] Furthermore, implantation of a conventional transvenous pacemaker in a young patient places them at risk for long-term device-related complications (Figure 3).[24] Therefore, a leadless system, especially in a patient such as ours with the primary conduction problem of AV block, is enticing. The Medtronic Micra™ leadless pacemaker was approved for use in the United States in 2016[25] and has shown excellent short-term success.[26] In paediatrics, small series with relatively short-term follow-up have been published.[27,28] Use of leadless pacing specifically for deglution syncope has been described in adults,[29] but not in paediatrics to date.

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Finally, when considering pacing, sensor-based systems such as closed-loop stimulation (Biotronik, Berlin, Germany) are a consideration.[30] Closed-loop stimulation has the potential to provide improved rate support during vagally mediated syncope, as it adjusts heart rate via sensing intracardiac impedance (reflecting myocardial contractility and autonomic tone), and thus potentially provides earlier chronotropic support than standard activity or ventilation-based sensors. For our patient, we ultimately favoured cardioneural ablation due to patient factors (young age) and the potential to directly attenuate the excessive parasympathetic input mediating syncope rather than merely compensate for the downstream effect.

Conclusion

While war, pestilence, famine, and death will always plague our planet, we need not tolerate a fifth Apocalyptic Horseman, namely, the inability to enjoy a delicious dish such as macaroni and cheese. Paediatric patients and their providers should harken to the news of improved treatment options for deglutition syncope, including cardioneural ablation and leadless pacing, that are now available when symptoms are refractory to less invasive therapy.

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Competing interests. None.

Ethical standard. Not applicable to this case report.

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