Anesthetic management of left ventricle non-compaction

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Introduction: Left ventricular noncompaction (LVNC) is currently categorized as an unclassified form of cardiomyopathy by the World Health Organization.(1) It is often mis- or undiagnosed due to limited awareness. Earlier diagnosis may allow modulation of therapy and improved outcome.

Case report: A 4 month old 6.3kg girl was noted to have sinus bradycardia (HR 40-50) at her regular visit to her pediatrician. She was transferred to our institution where a diagnosis of LVNC was made by echocardiographic examination. Despite her heart rate remaining between 50-80 at all times, she appeared in no distress and was hemodynamically stable. Additional echocardiographic findings confirmed good left ventricular function and a patent ductus arteriosus (PDA). She next presented to the operating room for permanent pacemaker placement and PDA ligation. Her vital signs in the OR were essentially unchanged with a heart rate of 60-70, arterial saturation of 99% in room air and normal blood pressure for her age. Two doses of atropine 0.1mg were administered which increased her heart rate into the 80 bpm range. Anesthesia was induced with etomidate and muscle relaxation was achieved using pancuronium. Anesthesia was maintained with isoflurane and incremental doses of fentanyl. She was stable throughout the operation and was transferred to the Intensive Care Unit where she was extubated on post operative day 1.

Discussion: LVNC is a relatively new diagnosis. Initially known as “spongy myocardium”, it was first reported in 1986 (2). It is thought to be caused by the persistence of the trabecular network of sponge-like muscle characteristic of mid to late embryonic life, when myocardial blood is supplied by diffusion from the intertrabecular spaces that communicate with the heart chambers. Diagnostic criteria are not yet universally defined but essential findings include 1) The presence of multiple echocardiographic trabeculations; 2) Multiple deep intertrabecular recesses communicating with the ventricular cavity; 3) A two-layer structure of the endocardium with a noncompacted to compacted ratio>1.4. (3,4). There does appear to be a genetic link although this again is yet to be elucidated. Our patient’s mother carried the same diagnosis and some paternal association is often seen. In addition, mutation of G.4.5 and alpha-dystrobrevin genes have been reported. Finally, skeletal muscle biopsy demonstrated mitochondrial morphological abnormalities and inclusions in some patients, suggesting a possible association of LVNC with underlying mitochondria myopathy (5). Patients typically present with congestive heart failure (CHF), ventricular arrythmias, or embolic events (although the latter may be rare in pediatric population). Previous reports suggested that the diagnosis carried a universally poor prognosis. However, although the natural history has not been definitively elucidated, it would appear that an early phase is followed by some period of remission, with relapse and worsening disease occurring in early adulthood. Long-term outcome is now thought to be more favorable is some subgroups(3).

Conclusion: This condition is under-diagnosed. Patients usually present with either congestive heart failure or with cardiac rhythm disturbances. Regardless of prior echocardiographic appearances, if this disease is suspected, a recent echocardiogram must be obtained prior to any procedure being undertaken. This is important because we know that in some individuals the course of the disease is variable. An excellent previous echocardiogram may have no bearing on current ventricular function. The potential association with mitochondrial myopathy should be remembered and appropriate precautions should be
taken. Finally, improved vigilance may lead to earlier diagnosis of cases, therapeutic interventions being initiated and possibly improve long term morbidity.

Reference

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