CASE REPORT

Does quality of life outweigh the cardiovascular risks of stimulant medication in a child with ADHD and hypertrophic cardiomyopathy?

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SUMMARY

A 10-year-old girl with attention-deficit hyperactivity disorder (ADHD) is diagnosed with hypertrophic cardiomyopathy. The stimulant medications used to control her ADHD pose possibly fatal risks to her cardiovascular health, so stimulant medication is stopped. Due to very poor quality of life off of medication, alternative therapies are used without improvement. The patient's caretakers decide that the benefits of stimulant medication outweigh the risks to the patient. The healthcare team clears the patient to be put back on stimulant medication with a signed waiver of liability by her caretakers.

BACKGROUND

Attention-deficit hyperactivity disorder (ADHD) is a chronic developmental disorder. It is described as a major clinical and public health problem because of its associated morbidity and disability in children. Children with ADHD are at risk for academic failure, emotional difficulties, poor relationships, and trouble with the law.¹ The use of stimulant medication is the most important variable in outcome.² Despite a historical black box warning, an updated safety review by the Food and Drug Administration (FDA) has reassured the public that there is no association with adverse cardiovascular events in healthy children. However, patients with 'serious heart problems, or for whom an increase in blood pressure or heart rate would be problematic'³ are advised not to be placed on ADHD medications. The medications studied included stimulants (amphetamine products and methylphenidate), atomoxetine and pemoline. An understanding of the physiological effects of stimulants on the cardiovascular system validates the concern despite a lack of data on clinical outcomes in this population. This relationship reveals an ethical dilemma as untreated ADHD can be emotionally crippling for a child and family. This case demonstrates that a well-informed family may choose the risk of stimulants over a compromised quality of life. How physicians address liability and manage patients on stimulants with cardiovascular disease is addressed in this case.

Case presentation

A 10-year-old girl presented to her paediatrician with episodes of concurrent palpitations and chest pain, which had been bothering her for a

few months. The episodes lasted 10 minutes and occurred on awakening and with exertion. She denied chills, lightheadedness, headaches, syncope, shortness of breath, or abdominal pain. She had no known allergies.

She was diagnosed with ADHD and had been taking Concerta for 4 years. She also had deficient visual-motor coordination, poor auditory processing and a learning disability. There was documented concern that she had a genetic abnormality linking her behavioural problems with her short stature and down-slanting palpebral fissures. This concern was not further investigated.

Her social history was complicated. Her biological mother and father used recreational drugs and alcohol during pregnancy. Her father died of a drug overdose before she was born and she was neglected by her mother in her first 3 years of life. When she was 4 years old, her mother's maternal rights were terminated and she was adopted by her great-grandparents.

She had no known family history of cardiovascular malformations.

On physical examination, she had a new grade 3 systolic murmur loudest along the left sternal border. She was in no acute distress and her vital signs were stable. She did not have jugular venous distension, abnormal breath sounds, hepatosplenomegaly, or peripheral oedema.

INVESTIGATIONS

Based on the patient's symptoms and new onset murmur, there was concern for possible myocarditis. She was referred to paediatric cardiology for evaluation. An ECG was obtained and was suggestive of left ventricular hypertrophy. An echocardiogram confirmed the diagnosis of concentric left ventricular hypertrophy with anterior displacement of the mitral anterior leaflet.

DIFFERENTIAL DIAGNOSIS

The diagnosis of hypertrophic cardiomyopathy (HCM) is identified when left ventricular hypertrophy is detected on echocardiography and there is no identifiable haemodynamic cause (eg, hypertension, valve disease).⁴ The underlying cause of HCM in a child is usually genetic. In this case, genetic testing for cardiomyopathies was advised. More specifically, her physical features and behavioural problems indicated testing for Noonan syndrome.

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There was concern that taking Concerta for 4 years may have contributed to or worsened the hypertrophy through haemodynamic mechanisms.

TREATMENT

The patient was advised to immediately stop taking Concerta due to the risk of sudden cardiac arrest and possible contribution to worsening hypertrophy. Her great-grandparents were hesitant to stop the medication due to the severity of her behaviour off of medication. The cardiologist and guardians agreed to stop taking Concerta with a follow-up in 4 months to see if the hypertrophy improved. Improvement would be assessed with a repeat echocardiogram.

There is no cure for HCM. Medical treatment with negative inotropic and chronotropic agents or placement of an implantable cardioverter-defibrillator is considered in patients who are symptomatic. The treatment is directed at reducing symptoms and sudden death.⁴

OUTCOME AND FOLLOW-UP

After 4 months of stopping Concerta, her behaviour dramatically worsened. She was struggling in school and at home. Her quality of life off medication was virtually unbearable for her guardians.

The degree of hypertrophy in her 4-month echocardiogram was unchanged. A 24-hour holter monitor did not show ventricular arrhythmias. The ethical dilemma weighing the quality of life benefits of stimulant usage against the risk of triggering potentially fatal dysrhythmias due to worsening her cardiomyopathy was difficult for her paediatrician, cardiologist, and psychiatrist to address. Given the lack of evidence-based medicine, there was an obvious concern for liability.

Her great-grandparents were informed and educated about the possible risks. They expressed their understanding and signed a waiver of liability stating that the prescribers were not responsible for poor outcomes. The patient was prescribed Concerta along with Atenolol and Clonidine for arrhythmia prophylaxis. She would have regular follow-ups every 6 months with her cardiologist that included echocardiography and holter testing as needed.

One month after beginning Concerta, the patient's great-grandparents felt that she was much more manageable. She was doing remarkably better in school, seemed dramatically happier, and was easier to communicate with.

Genetic testing with high-resolution microarray-based comparative genomic hybridisation identified a 1.3 Mb duplication of 16p13.11. The 16p13.11 duplication is characterised by predominant skeletal features, cardiac and aortic malformations, cognitive impairment and behavioural abnormalities including ADHD.

Discussion

The management of ADHD in children with congenital heart conditions is a serious ethical dilemma with unclear guidelines to direct physicians. The FDA does not recommend prescribing stimulants to patients in whom an increase in blood pressure or heart rate would be problematic. However, if ADHD therapy is 'deemed necessary',³ it should be managed. What is considered 'necessary' is not clearly defined, making prescribing physicians uncomfortable. There have been no similar publications that discuss this dilemma and the liability of prescribing physicians.

A case report titled 'Liability Associated with Prescribing Medications'⁵ defines how physician liability is determined in the legal

arena. Liability for unintentional harm requires the presence of 'The Four Ds' of malpractice: The physician has undertaken the patient's treatment and has a 'duty' to the patient. The physician is 'derelict' in this duty which 'directly' causes the 'damage' or harm to the patient. Determining when a physician is derelict, or not practising the standard of care, is relevant to this case because the guidelines for ADHD treatment in children with HCM are poorly defined. The report emphasises that physicians are protected from dereliction of duty when the standard of care is met in the form of informed consent.⁵

The medical community agrees that despite the utility of signed consent forms in the legal arena, it should remain an educational model for patients and not a waiver of liability for physicians. When informed consent forms are a template generated by attorneys and not for the individual patient, the educational model is lost. Informed consent forms should be personalised and not template-generated. They should serve as a documentation of discussion. In this case, the child's great-grandparents were educated about the risks of continuing stimulant medications despite her cardiovascular condition. They agreed to sign a waiver of liability to document that they fully understood the risks and wanted to move forward with stimulants despite the risks.⁶

Learning points

- ▶ Due to the risks associated with stimulant medication and hypertrophic cardiomyopathy, children with attention-deficit/hyperactivity disorder (ADHD) and hypertrophic cardiomyopathy should attempt a trial off of stimulant medications.
- ▶ If quality of life is intolerable off stimulant medication, physicians should educate the family on the risks of resuming medication with appropriate documentation.
- ▶ Clinical research is necessary to determine the safety of stimulant medications in children with ADHD and hypertrophic cardiomyopathy.

Contributors All persons who meet authorship criteria are listed as authors, and all authors certify that they have participated sufficiently in the work to take public responsibility for the content. These authors include: ES (primary author), JS (secondary author) and MS (attending physician). Furthermore, each author certifies that this material or similar material has not been and will not be submitted to or published in any other publication before its appearance in the BMJ case reports. This case report is written about a pediatric patient of MS. The ethical dilemma in handling this patient's care stimulated discussion among us. This led to a thorough search for guidelines on patient management and a literature search to identify similar cases. We believed that the discussion and treatment of this patient was worthy of a publication. ES and JS wrote the case report in full disclosure with the family and doctors involved. MS mentored us through the process. There were no data collected for this submission.

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