No heart block without a cause

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To the editor:

We read with interest the article by Gutierez-Gallego et al. about an 11 years-old female who initially manifested with non-migrainous headache and exercise-induced syncope [1]. Initial work-up revealed an AV-block-III in the absence of any comorbidity and extensive work-up did not detect an underlying cause [1]. According to the guidelines, the patient neither received an antiarrhythmic medication nor a pacemaker [1]. The report is promising but raises concerns that should be discussed.

We disagree with the classification of AV-block-III as idiopathic [1]. There is no disease without a cause. It usually depends on the investigational methods applied for the work-up of disease and on the meticulousness of the work-up if the underlying etiology is discovered or not. Regarding the index case, work-up for the cause of the AV-block-III could have been more extensive.

Work for the cause of AV-block-III was incomplete with regard to a putative genetic cause. Not only Kearns-Sayre syndrome may manifest with an AV-block-III but also several other syndromic and non-syndromic mitochondrial disorders [2]. Since the family history was not provided in detail, it cannot be excluded that the index patient had inherited a mitochondrial disorder from either parent. Whole exome sequencing is missing.

The patient was diagnosed with dysautonomia but it is not mentioned upon which criteria dysautonomia was diagnosed. Did the patient undergo a tilt-test investigation, frequency analysis of Holter monitoring, or a skin biopsy? Were there any other clinical manifestation of autonomic neuropathy, such as oversensitivity to light, dry mouth, obstipation, or urinary dysfunction? It is crucial that small fiber neuropathy is adequately ruled in the index patient.

The reason why the patient initially manifested with headache has not been clarified. However, it can be speculated that headache as the initial manifestation of an AV-block-III could be due to cerebral hypoxia due to impaired cerebral perfusion. It is also conceivable that the patient experienced cerebral vasospasms, as in migraine attacks. Third, it cannot be definitively ruled out the patient had experienced a venous sinus thrombosis due to the hemodynamic changes triggered by the AV-block-III.

It is not comprehensible why the patient was diagnosed with migraine, although the headache's character was described as non-pulsatile, there was no photophobia, and no autonomic dysfunction such as nausea, emesis, or hypothyroidism.

We wonder how the clinical neurologic exam can be normal given the fact that the patient had dysautonomia. Since the patient was admitted for syncope it is crucial that a cerebral cause of the syncope is ruled out. We should be informed about the results of the cerebral magnetic resonance imaging, carotid ultrasound, and the electroencephalography. Were there any indications for anemia or even pancytopenia suggesting Pearson syndrome, a precursor of Kearns-Sayre syndrome?

We disagree with the notion that the prognosis of idiopathic AV-block-III is generally good, as mentioned in the discussion [1]. The outcome of an AV-block-III depends on the underlying cause of the AV-block-III and may range between favourable and death.

Overall, the interesting study has some limitations that call the results and their interpretation into question. Clarifying these weaknesses would strengthen the conclusions and could improve the study. In pediatric as well as adult patients presenting with a new AV-block-III extensive work-up should be initiated until the underlying cause has been revealed.

Notes

Contributor roles

JF: design, literature search, discussion, first draft, critical comments, final approval. SM: literature search, discussion, critical comments, final approval.

Competing interests

The author declares that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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References

1. Gutiérrez-Gallego JM, Estrada-Perez MF, Barrios-Arroyave FA, Torrente-López JD, Correal-Barrios A. High-grade idiopathic atrioventricular block in childhood: Case report and literature review. Medwave. 2022;22. https://doi.org/10.5867/medwave.2022.05.002535
2. Niedermayr K, Pölzl G, Scholl-Bürgi S, Fauth C, Schweigmann U, Haberlandt E, et al. Mitochondrial DNA mutation "m.3243A>G"-Heterogeneous clinical picture for cardiologists ("m.3243A>G": A phenotypic chameleon). Congenit Heart Dis. 2018;13: 671–677. https://doi.org/10.1111/chd.12634