

Intracardiac Yolk Sac Tumor and Dysrhythmia as an Etiology of Pediatric Syncope

Melinda J. Morin, MD*; Richard A. Hopkins, MD‡; William S. Ferguson, MD§; and James W. Ziegler, MD||

ABSTRACT. This report describes a new etiology of pediatric syncope. Epilepsy, brain anomalies, infection, electrolyte abnormalities, and trauma are commonly identified etiologies of seizures in the pediatric population. We report here a child with third-degree heart block and right ventricular outflow tract obstruction related to an intracardiac tumor presenting with syncope and seizure-like activity. Echocardiography revealed a large (3 × 8-cm) intracardiac mass filling the right atrium, extending across the tricuspid valve into the right ventricle and crossing the atrial septum into the left atrium, extending into the left ventricular outflow tract. She underwent emergent cardiopulmonary bypass with removal of the majority of the tumor mass, clearing both the left and right ventricular outflow tracts of obstruction and repairing the tricuspid valve. Postoperative cardiac conduction remained blocked and required permanent pacing. The initial serum α -fetoprotein level was grossly elevated, and the tumor showed characteristic histopathologic features of a yolk sac tumor. Four years after the completion of her chemotherapy, she remains clinically well, with no evidence of recurrent tumor by echocardiography or radiographic studies, and her α -fetoprotein remains in the normal range. The clinical manifestations of tumor infiltration of the heart with complete heart block resulting in loss of consciousness with tonic-clonic movements are detailed. Although rare, cardiac syncope has multiple known causes and should be suspected in any patient with sudden loss of consciousness and pallor. In the pediatric population, cardiac rhythm disturbances are typically the result, rather than the cause, of acute cardiac emergencies. Pediatricians should be aware of depressed cardiac output and dysrhythmias as etiologies of new-onset syncope. Evaluation should include a cardiac assessment with electrocardiogram to exclude a life-threatening arrhythmia as a potential cause. *Pediatrics* 2004;113:e374–e376. URL: <http://www.pediatrics.org/cgi/content/full/113/4/e374>; *yolk sac tumor, dysrhythmia, seizure*.

ABBREVIATIONS. A-V, atrioventricular; ECG, electrocardiogram; AFP, α -fetoprotein.

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Seizures are a common reason for pediatric visits to an emergency department. Epilepsy, brain anomalies, infection, electrolyte abnormalities, and trauma are commonly identified etiologies of seizures in the pediatric population.^{1–5} Cardiac dysfunction resulting in inadequate delivery of oxygen and metabolic substrates to the brain is a less common cause of altered consciousness with seizure-like movements. The clinical symptoms of cerebral hypoperfusion depend on the severity and duration of the ischemia. Sudden, severe loss of cerebral perfusion can result in loss of consciousness and seizure activity. We report here a child with third-degree heart block and right ventricular outflow tract obstruction related to an intracardiac tumor presenting with syncope and seizure-like activity.

CASE REPORT

A 3-year-old previously healthy girl presented to an outside hospital with a history of a transient loss of consciousness while on the toilet, with subsequent tonic-clonic movements. Initial evaluation revealed a well-appearing, communicative child with no cervical spine injury by radiologic examination. She was discharged from the hospital, but several hours later, she again experienced loss of consciousness and was transported by ambulance to our tertiary care medical center. Of note, she had no previous history of alterations in mental status or seizure-like activity. There was no history of fever, rash, insect bites, or changes in weight or activity level.

On physical examination, she was observed to have multiple, brief episodes of pallor associated with loss of consciousness and generalized tonic-clonic activity, between which she was well appearing. She was afebrile. Between the episodes of loss of consciousness, her heart rate was 90, respiratory rate was 30, and blood pressure was 130/80. Her cardiac examination was notable for a grade II/VI pulmonic ejection murmur and a pulse that varied between regular and irregularly irregular with brisk capillary refill. There was no jugular-venous distension or increased work of breathing. She had clear breath sounds and no hepatosplenomegaly. She had equally round and reactive pupils and a grossly nonfocal neurologic examination. A rhythm strip was obtained, which demonstrated complete atrioventricular (A-V) dissociation with a narrow complex escape rhythm. Her “spells” were associated with 15- to 20-second episodes of ventricular asystole. The patient was transferred to the pediatric intensive care unit.

An electrocardiogram (ECG) (Fig. 1) revealed third-degree heart block with a junctional escape rhythm at 80 beats per minute and episodes of ventricular asystole of 15- to 20-second duration with nonconducted p waves. Echocardiography revealed a large (3 × 8-cm) intracardiac mass filling the right atrium and extending across the tricuspid valve into the right ventricle. Moderate right ventricular inflow and outflow obstruction was present. The mass crossed the atrial septum into the left atrium and extended into the left ventricular outflow tract. The free walls of the cardiac chambers were not involved.

She was stabilized with mechanical ventilation and infusions of epinephrine and atropine and was brought to the operating room. She underwent emergent cardiopulmonary bypass with removal

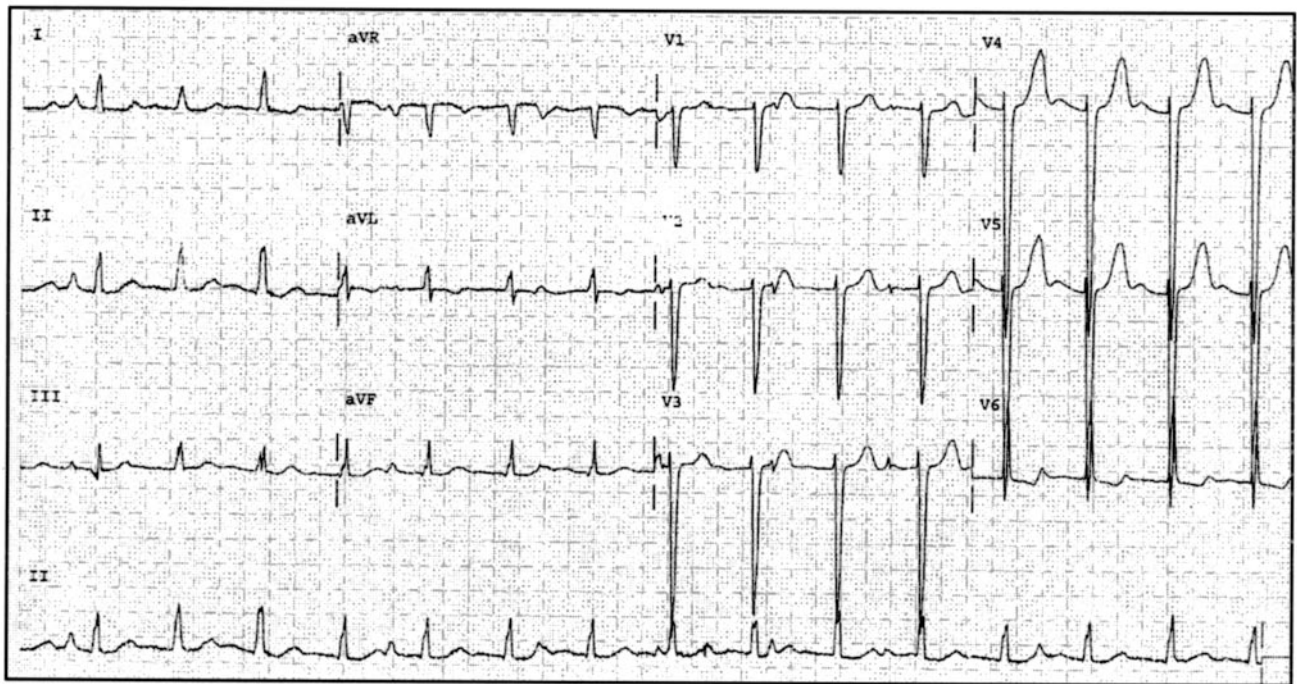


Fig 1. Presenting ECG (asymptomatic) demonstrates A-V dissociation with a regular escape rate of 90 beats per minute.

of the majority of the tumor mass, clearing both the left and right ventricular outflow tracts of obstruction and repairing the tricuspid valve. The atrial septum was removed along with infiltrated tumor tissue and replaced with patch material, but remnants of the tumor, which appeared to originate from the central fibrous body, were left so as not to disrupt mitral or tricuspid valve functions. In addition, complete removal would have resulted in a large ventricular septal defect and the need for prosthetic replacement of both A-V valves. The septal tricuspid valve leaflet was reattached to the pericardium and plicated to the anterior leaflet. Because of existing heart block, both temporary and permanent epicardial pacing leads were attached to the heart. The epicardial pacing leads were buried in the subcutaneous pocket in the right upper quadrant of the abdomen for later retrieval.

Her postoperative course was uncomplicated, and she was extubated 12 hours later. Echocardiography demonstrated no shunt across the ventricular or atrial septum and normal mitral valve function. There was mild tricuspid valve regurgitation along the septal region where the septal leaflet had been displaced and infiltrated by the tumor requiring valve repair. Cardiac conduction remained blocked (likely because of tumor infiltration and destruction of the A-V node). Therefore, 4 days later, a permanent pacemaker was placed in the previously prepared subcutaneous upper abdominal pocket and attached to the previously positioned epicardial leads.

The tumor showed characteristic histopathologic features of a yolk sac tumor including myxoid, papillary, solid patterns, and Schiller-Duval bodies.⁶ Immunohistochemical staining was supportive with positive cytokeratin, placental alkaline phosphatase, and focal α -fetoprotein (AFP) staining. Variability in AFP immunostaining is well described and may depend on the histologic component as well as the laboratory or antibody in use. In our experience, the serum levels correlate much better than immunohistochemical staining. This also applies to our experience with hepatocellular carcinoma. The lack of human chorionic gonadotropin-positive syncytial giant cells argues for the solid component of yolk sac tumor rather than an embryonal carcinoma component. Lack of immature ectodermal and mesodermal components, in particular, immature neural tissue, precludes a diagnosis of immature teratoma.⁷ Her initial serum AFP level was 12 560 ng/ml (normal: <10 ng/ml); there was no radiographic evidence of metastatic disease.

Chemotherapy was initiated 1 week after the debulking procedure. The patient received a total of four 5-day cycles of BEP (bleomycin, 15 U/m² on day 1; etoposide, 100 mg/m² on days 1-5;

and cisplatin, 20 mg/m² on days 1-5). Four years after the completion of her therapy, she remains clinically well with normal growth, development, and activity levels. She has no cardiac symptoms, and her pacemaker continues to function well. She is doing well in school and exhibits no neurologic or intellectual deficits. There is no evidence for recurrent tumor by echocardiography or radiographic studies, and her AFP remains in the normal range.

DISCUSSION

More than stroke volume, heart rate is the primary determinant of cardiac output in infants and children.⁸ In this case, the cardiac conducting system of this child was ablated by tumor and, as the period of time between episodes of ventricular standstill shortened, she became increasingly symptomatic. It is likely that the patients' escape junctional rhythm was regular during her initial emergency department visit, because no ECG was performed and a cardiac etiology of her symptoms was not entertained. Hemodynamic compromise was not appreciated as the cause of her symptoms until the combination of pallor and loss of consciousness prompted initiation of cardiac monitoring. In the absence of the classic symptoms of poor cardiac output (poor urine output and feeding, decreased activity, cyanosis), cardiac dysfunction in children is rarely suspected.

Clues suggesting that this patient's symptoms were cardiogenic and not neurologic in origin include the absence of any aura preceding loss of consciousness, the marked pallor noted during spells, and the rapid return to normal mental status with no postictal state. Syncope from any cause results from cerebral hypoperfusion and can be associated with seizure-like activity, often making the distinction between syncope and seizures difficult. Although autonomic dysfunction, manifested as pallid breath-holding spells early in life and vasovagal syncope

later in childhood, is the most common cause of syncope during the pediatric years, this patient had no stimulus or stress provoking her episodes. In addition, these episodes occurred in the recumbent position. Although rare, cardiac syncope has multiple known causes and should be suspected in any patient with sudden loss of consciousness and pallor.

Cardiac tumors are rare in children, and tumor location plays an important role in the clinical presentation. Patients often display symptoms of cardiac failure, arrhythmias, valve dysfunction, and pericardial effusions.^{9–12} This child's presenting symptoms of syncope and seizure-like activity are typical of Stokes-Adams attacks,¹³ commonly seen in patients with complete heart block. Failure of an adequate escape rhythm results in sudden loss of cardiac output. Seizure activity caused by this mechanism has been reported in adults¹⁴ and children.¹⁵ In the pediatric population, cardiac rhythm disturbances are typically the result, rather than the cause, of acute cardiac emergencies. However, cardiac rhythm abnormalities must be suspected in any patient presenting with atypical syncope.

Germ cell tumors are rare in the pediatric population, comprising only ~1% of childhood tumors. Approximately 50% will occur in extragonadal sites, although the majority of these will be sacrococcygeal teratomas. Of the malignant extragonadal germ cell tumors, a yolk sac component will be present in up to 60% of patients.^{16–19} Although historically the prognosis for malignant germ cell tumors was poor, with platinum-based multiagent chemotherapy the overall cure rate in children for extragonadal germ cell tumors is ~80%, and for gonadal tumors the cure rate approaches 90%.²⁰

CONCLUSIONS

We report the first case of an intracardiac yolk sac tumor presenting with syncope and seizure activity in a preschool-aged child. The possibility of diminished cardiac output and cardiac dysrhythmias should be kept in mind when evaluating a child with sudden loss of consciousness. Rapid cardiopulmonary assessment using the ABC (airway, breathing, circulation) approach advocated by the American Heart Association²¹ should be performed on all ill pediatric patients to determine the severity of physiologic derangements. In the absence of an obvious explanation for a syncopal event, the evaluation should include a cardiac assessment with ECG to

exclude a life-threatening arrhythmia as a potential cause.

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