A case of an anomalous origin of left main coronary artery from right sinus of valsalva leading to sudden death

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We spend the bulk of our efforts attempting to understand the basic framework of medicine and the top 5-10 entries on differential lists. And for good reason – these are the things that we will assuredly see and have to treat. There are always, however, going to be cases that fall outside of the realm of the usual. These cases puzzle the best amongst our profession and make us wonder if there was anything we could have done differently.

A young adolescent female presented to a pediatric emergency department with chest pain, shortness of breath, and cyanosis. She had been jogging when the chest pain started, and then collapsed. Except for a previous possible seizure or syncopal episode a few years earlier, she was otherwise healthy. Initial vital signs were a heart rate of 85 bpm, blood pressure of 70/40 mmHg, respiratory rate of 24 breaths/min, temperature of 37°C, and oxygen saturation of 68% on room air. An electrocardiogram displayed ST changes suggestive of ischemia.

The initial presentation suggests that she was in shock – a pathophysiologic state of inadequate tissue perfusion and subsequent hypoxia. It is subcategorized by the underlying mechanisms: reduced blood volume (“hypovolemic shock”), a pathological distribution of blood volume (“distributive shock”), and/or failure of the pump (“cardiogenic pump”). If corrected quickly, the short-term effects of poor perfusion are generally reversible. Prolonged shock, however, will eventually lead to irreversible damage including cell death, end-organ damage/failure, and death.

Typically, pediatric patients presenting with shock in the emergency department are experiencing hypovolemic shock due to dehydration (resulting from diarrhea or osmotic diuresis) or trauma (and subsequent hemorrhage), or distributive shock from sepsis. The routine work-up for such a patient is to identify and address any life-threatening conditions (via a rapid assessment of the patient’s general appearance, breathing quality and rate, and circulation to skin), recognize circulatory compromise, and then identify the type of shock and find the underlying cause. The detective work involved with this last step includes a detailed history and physical exam, combined with the necessary ancillary tests.

Our patient’s history and physical findings suggested a cardiogenic cause of shock. One of the more alarming aspects of her presentation was her low blood pressure and heart rate. In pediatric populations, a low blood pressure indicates a later and more severe stage of shock. Clinically, shock has been described in three progressive stages. The first stage is compensated shock, in which the body’s homeostatic mechanisms (increased heart rate and peripheral vasoconstriction) are able to maintain adequate perfusion and systolic blood pressure. If left untreated, compensated shock progresses to hypotensive shock, in which the compensatory mechanisms are overwhelmed and there is a subsequent drop in systolic blood pressure along with signs of poor perfusion (e.g. altered mental status). If the inadequate organ perfusion continues, a patient will eventually progress to irreversible shock, in which end organs are irreparably damaged, and despite resuscitation, the patient will die.

To prevent the progression through these stages of shock, current guidelines advise an aggressive, multi-pronged treatment algorithm aimed at improving perfusion and end organ function. The first arm is rapid fluid resuscitation using isotonic crystalloid solution. Early vascular access is needed to facilitate fluid resuscitation efforts. Another treatment arm is airway and respiratory support in the form of supplemental oxygen, positive pressure ventilation, and/or intubation. Monitoring of physiological indicators (including blood pressure, quality of pulses, skin perfusion, mental status, and renal output) before and after interventions provide additional clues to the type and underlying causes of shock. This information would direct the final arm of treatment, the selection of appropriate medications.

Our patient exhausted the available treatment modalities. She was started with peripheral venous lines. Gravity alone was not enough to push the requisite amount of fluid. Direct pressure on the bag and “push-pull” method were employed to maximize fluid delivery. Additional vascular lines were established using bilateral intraosseous (IO) infusions into her proximal tibias. These infusions take advantage of the veins that drain the medullary sinus of long bones. Due to their support from the bony-matrix, these veins do not collapse. After cannulation to gain access to the medullary sinus using a large bore needle and drill, and flushing with normal saline, an IO has equivalent infusion rates to a 21 gauge peripheral intravenous catheter. She was placed on supplemental oxygen, intubated, and given inotropic medications (dopamine at 5-20 mcg/kg/min, norepinephrine at 0.05-0.5 mcg/kg/min, and epinephrine at 0.1-3.0 mcg/kg/min). Her cardiac output deteriorated, and eventually resuscitation attempts were discontinued.

The post-mortem examination of the heart revealed an anomalous origin of the left main coronary artery (LMCA) from the right sinus of...
Valsalva. The precipitating cause of death for our patient was determined to be acute myocardial ischemia. The standard shock management regimen was not sufficient to treat her condition, and parts of the administered therapy may have been detrimental.

Under normal circumstances, there are two sinuses of Valsalva arising from the aorta distal to the aortic valve. Anomalous origin of coronary arteries (AOCA) is a rare (estimated 0.1-0.3% prevalence) congenital heart defect, in which coronary vessel(s) follow an abnormal route (see Figure 1). Our patient’s left main coronary artery arose from the right sinus, passed between the pulmonary trunk and ascending aorta, before supplying its dependent heart tissue. Despite its low prevalence, AOCA is the second leading cause of sudden death in young athletes, representing approximately 10% of deaths in this population. This discrepancy may be due to a high mortality rate associated with this condition and/or the under diagnosis of this condition (affected individuals are typically asymptomatic during daily activities of living).

AOCA is often included in the differential for sudden cardiac deaths in young – albeit as a diagnosis of exclusion. Even so, when the condition actually causes a coronary event due to the insufficient oxygen delivery when placed under stress, its presentation is very difficult to discern from other conditions that cause similar symptoms. Most cases are identified upon autopsy, where myocardial fibrosis in the chronically under-perfused heart tissue and signs of hyper-acute myocardial infarction may be evident. In our case, the young girl was given inotropic agents in order to increase her cardiac output by increasing force of ventricular contraction. However, unbeknownst to the physicians, these agents further enlarged her pulmonary trunk and aorta upon each heartbeat, further occluding her LMCA, and thus exacerbating the ischemia of the heart muscle.

A case like our patient’s will always beg the question, what could have been done differently? The reality of this case is that AOCA is incredibly difficult to identify, especially in the emergency setting. However, there is always something that can be gained from evaluating any and all aspects of the case in order to gain wisdom and knowledge to aid us in future patient care. In this case, there were a few clues. The first clue was her history. From the literature, the myocardial infarction in individuals with AOCA is often preceded by a period of strenuous exercise. Our patient was in her teens, otherwise healthy, and had been jogging before experiencing non-specific chest pain, followed by collapse and loss of consciousness. Exertional syncope is always more concerning than non-exertional. In conjunction with her response to treatment, this presentation may have raised suspicion of a myriad of cardiac conditions, including AOCA. More importantly, on further questioning, the initial presentation had been precipitated by exertion. She had fully recovered from that incident, but if she had been sent for further evaluation it is possible that the defect may have been detected before she had a second syncopal episode, and may have been surgically corrected.

Heart function tests including the ECGs and ejection fractions that were done for our patient have not been demonstrated to be helpful diagnostic tools for coronary artery anomalies. Imaging, however, allows physicians to note the presence of a congenital defect in the origin of the coronary, follow the artery’s unusual course, and assess the integrity of the artery by looking for regions of stenosis. One of the best methods to identify this anomaly is the use of computed tomographic angiography (CTA) to visualize the displaced coronary artery. This type of imaging is unfortunately not practical in such an emergency setting. Transthoracic and transesophageal echocardiography have also been demonstrated to be feasible and practical in young patients, and these imaging modalities can be used to justify ordering a CTA scan. Our patient did get an emergent echocardiogram during active resuscitation, but the vessels were not clearly visualized.

Upon identification of the anomaly, treatment is possible through surgical means. Coronary artery bypass grafting (CABG) remains the standard treatment in these instances, with some surgical teams electing to also ‘unroof’ the associated anomalous coronary artery. This technique involves modifying the ostium of the coronary artery by excising (‘un-roofing’) the common wall between the aorta and anomalous artery. Attempts to re-implant the anomalous vessel into the correct aortic sinus have been completed, but are technically challenging and are not yet indicated over the safer surgical alternatives. Our patient was never stable enough to consider this option.

The medical profession is ripe with challenges for practicing physicians. One of the most mentally difficult tasks required of physicians is the need to maintain an expert knowledge of countless medical conditions, many of which are exceedingly uncommon epidemiologically. Losing a patient who could have been helped by the correct treatment if the proverbial “zebras” had been recognized in time is a true test of a physician’s confidence, and is a stark reminder that the science and art of medicine demands perfection from imperfect people.

REFERENCES