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Letters to the Editor Methemoglobinemia: Arterial blood gas as a diagnostic tool



Methemoglobinemia is an important differential diagnosis of a case of cyanosis with no underlying structural heart disease. We report 2 cases of this rare disorder who were referred to our hospital with cyanosis.

1. Case 1

A 17-day-old male child was referred with dusky extremities noticed few days after birth and suspected to have cyanotic congenital heart disease. His basal oxygen saturation was 79%. Rest of the respiratory and cardiac examination were unremarkable. Echocardiography revealed no underlying structural heart disease. Arterial blood gas (ABG) revealed PO2 = 285 mm Hg with elevated MetHb levels (44%). On oxygen supplementation there was increase in PO2 to 400 mm Hg but SPO2 remained at 90%. Filter paper test done revealed significant discoloration (compared to normal). His electrophoresis and G6PD (Glucose 6 Phosphate dehydrogenase) levels were done. The baby was given intravenous methylene blue and ABG was repeated after 30 min which revealed a significant reduction in the methemoglobin levels (1.7%). Repeat ABG after 12 h showed methemoglobin level of 3.9%. Simultaneously he was also started on Vitamin C and riboflavin. Parents ABG analysis documented normal methemoglobin levels and normal SPO2. The patient was subsequently discharged with stable vitals.

2. Case 2

A 32-year-old non hypertensive male presented with complaint of sudden onset cyanosis noticed since 1 day. On evaluation he had history of fever for 5 days and was advised chloroquine empirically. On examination he had stable vitals with normal heart sounds and no murmur. 2-dimensional echocardiography was within normal limits. Blood investigations including liver and renal function tests were within normal limits. His methHb level was found to be high (30%) on arterial blood gas analysis. His arterial oxygen saturation showed gradual improvement from 86% to 92%. He was managed conservatively. He was subsequently discharged on 3rd day and is doing fine on follow up with no recurrence of cyanosis.

3. Discussion

Cyanosis with structurally normal heart is always a matter of great dilemma. Methemoglobinemia is an important differential diagnosis whenever thinking of an acquired or drug induced cause. Hemoglobin can accept and transport oxygen only when the iron atom is in its ferrous form. When hemoglobin loses an electron and becomes oxidized, the iron atom is converted to the ferric state (Fe³⁺), resulting in the formation of methemoglobin. Methemoglobin lacks the electron that is needed to form a bond with oxygen and thus is incapable of oxygen transport. The low level of methemoglobin is maintained through 2 important mechanisms. The first is the hexose-monophosphate shunt pathway within the erythrocyte. Through this pathway, oxidizing agents are reduced by glutathione. The second and more important mechanism involves 2 enzyme systems, diaphorase I and diaphorase II, and requires nicotinamide adenine dinucleotide (NADH) and nicotinamide adenine dinucleotide phosphate (NADPH), respectively, to reduce methemoglobin to its original ferrous state. Any drug that interferes with these mechanisms can lead to methemoglobinemia.^{1,2} Thus, it is important to take an appropriate drug history particularly for drugs like chloroquine as was observed in our second case.^{1,2} Acquired methemoglobinemia incidence may be much higher than is documented. Often, this is associated with the use of or exposure to oxidant drugs, chemicals, or toxins, including dapsone, local anesthetic agents, and nitroglycerin. Arterial blood with elevated methemoglobin levels has a characteristic chocolate-brown color as seen in our first case. A normal arterial blood gas analysis machine does give the value of methemoglobin and shows high po2 levels in the presence of cyanosis. Both the above mentioned cases highlighted these findings as methHb levels were overlooked in the routine arterial blood gases by the referring center. A low oxygen saturation by pulse oximetry measured in patients with normal arterial blood gases can be an indication of methemoglobinemia.³ A few ABG machines do give a value of methhemoglobin levels too. In the absence of routine habit to look keenly at the value, the diagnosis of the methemoglobinemia is likely to be missed, the same happened with our 2nd case when he was referred to us by the referring center. In most

cases of mild acquired methemoglobinemia, no treatment is needed. Prompt recognition of the condition and initiation of treatment, as indicated (especially in acquired methemoglobinemia), are critical in the management of methemoglobinemia for higher levels of methemoglobin levels. Intravenous (IV) methylene blue is the first-line antidotal agent. Exchange transfusion and hyperbaric oxygen treatment are second-line options for patients with severe methemoglobinemia whose condition does not respond to methylene blue or who cannot be treated with methylene blue (e.g. with g6pd deficiency).

To conclude arterial blood gas may be a useful indicator for methemoglobinemia and should be analyzed for methemoglobin levels in an undiagnosed case of cyanotic heart disease with structurally normal heart.

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Integrating spirituality into patient care: An essential element of modern healthcare system

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World Health Organisation has already realized the need of the 4th dimension of health, i.e. the spiritual health to be considered as an important element of health. In the words of Derek Yach (World Health Assembly May, 1998):"From the inception, it was felt that the 4th Dimension of health was missing from its definition. The special group of the WHO Executive Board (1998) proposed that the Preamble of the Constitution should be amended as follows":

"Health is a dynamic state of complete physical, mental, spiritual and social well being and not merely the absence of disease or infirmity."¹

During one of my lectures on "Life style modification and Health" I was asked to describe spiritual health. Although spiritual health means something different to everyone, below is my response hoping that it might offer some insight in addition to an expanded definition of health beyond diet and exercise. Again, this is my own perspective on spiritual health, which is a dynamic and ever evolving field of learning in medical science and hence not to be considered as absolute truth.

The traditional meaning of spirituality is a process of reformation which "aims to recover the original shape of man, the image of God". One can define spiritual health as nothing but peacefulness, simplicity, empathy, compassion to name a few. It might really sound to be true and interesting. It does happen with me too, whenever, I am around a spiritually healthy person, I feel peaceful, inspired, relaxed and safe. It is said, a spiritually healthy person is very much in tune with the present moment and doesn't live in the past or in the future, but instead fully accepts the current moment as the only "real" moment in which to experience and enjoy life in totality.

Interestingly, spirituality and health is a growing field of healthcare. It grew out of courses in spirituality and health developed for medical students in the United States. Research in this area over the last 30 years has also formed an evidence base for spirituality and health. Studies have demonstrated an association between spiritual beliefs of patients and values and a variety of healthcare outcomes. More recent research has also shown a strong desire on the part of patients to have their spirituality addressed as part of their medical care.

Empirical evidence is also available to indicate a direct relation between spirituality and positive health outcomes. Positive values, attitudes, belief and strength that one acquires through spiritual practices contribute to health and happiness. Spiritual practices have a positive correlation with survival, low blood pressure, fewer symptoms of anxiety and