Incidental Diagnosis Of Methemoglobinemia In Patient For Atrial Septal Defect Closure: Case Report

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Abstract

Abnormal hemoglobin or hemoglobinopathies are not uncommon in India. Most of them can be diagnosed on photo spectrometric studies rather than symptoms or signs alone.1,2 Incidental finding such as “Dark color arterial blood” with low SPO2 on pulse oximetry can raise suspicion of Hemoglobinopathies or abnormal Hemoglobin. It can be further confirmed on investigations.

This is a case report of a patient who was having “Dark blood”, low Spo2 not responding to increase in FiO2 with normal PaO2 levels: operated for Sinus Venosus Atrial Septal Defect (ASD). He was diagnosed to have high levels of Meth-hemoglobin on further Investigations.

CASE REPORT

A 20 years male, known case of Sinus venosus ASD since childhood presented with history of easy fatigability since 3-4 months. His previous, personal and family history were insignificant.

On examination, Pulse Rate was 80/min., BP 110/70 mmHg, without evidence of cyanosis or clubbing. On auscultation of chest, S1S2 were normal without any murmur. Air entry equal and clear on both sides.

On Investigations, Hb was 16.8gm%, Rest of hematological investigations were within normal range. Pre-op echocardiography was showing large sinus venosus ASD with left to right shunt: Right upper pulmonary vein draining to Right Atrium (PAPVC), grade II Tricuspid Regurgitation. Moderate pulmonary hypertension. Left superior vena cava with dilated coronary sinus. Ejection Fraction 55%.

Patient was scheduled for surgical closure of atrial septal defect. After overnight fast & premedication with tab. Diazepam, tab. Pantoprazole the patient was trolleyed to Operating Room.

Standard monitors including ECG, Pulse-oximetry, non-invasive blood pressure cuff were attached. Patient was in sinus Rhythm. Pulse oximetry revealed the saturation 90% on room air. Oxygen at a flow rate of 8L/min was started through Hudson’s face mask. Saturation was 94% after 3mins of oxygenation with Face Mask and remained the same with gradual increase of oxygen flow upto 10 li/min.

Under local anesthesia, (Inj. Lignocaine 2%, 1cc), intravenous access was secured. Sedation 1mg Inj. Midazolam and Inj. Fentanyl 50 mcg. given intravenously. Antibiotic was administered.

Under local anesthesia (Inj. Lignocaine 2%, 2cc) Right radial artery was cannulated. The unusual dark colour of blood was confusing for a vein. But jet and flow was like arterial puncture. Further arterial cannulation was confirmed on pressure transducer by Pressure of 140/80mm Hg. Arterial blood gas analysis showed Pao2 239 on face mask O2 flow 8li/min. saturation 99.5% and no acidosis. But Pulse oximerty was showing SpO2 of 94%.

Anesthesia was administered with Inj. Fentanyl 300mcg, Inj. Midazolam 3mg., inj. Pancurnium 8mg. The trachea was Intubated with 8.5 no. PVC cuffed ETT.

Maintenance of anesthesia was done with oxygen and Isoflurane gas mixture. Left sided IJV was cannulated with 7 no. Triple lumen.

Surgical procedure was started, after adequate

heparinazation Aortic and Caval cannulation was done. The color of blood column seen in both lines was almost same and was Chocolate Red Color. On CPB Pump Pao2 was normal for FiO2 without any acidosis.

Post-surgery, CP Bypass was commenced uneventfully with ionotropic support Inj. Dobutamine 5mcg/kg./min. Post Bypass SpO2 was same as preop ie. 94% with normal PaO2.

Table 1

Blood Gas Analysis showing Values of Oxygen Saturation in blood (PaO2), Oxygen saturation on Puse oximetry (SpO2) at different FiO2 levels.

	FiO 2	PaO2%	PaO2 mmHg	SpO2%	PaO2-SpO2 **Saturation Gap**	Base Deficit
Room air	0.21	-		90	-	-
Face Mask	0.4	99.5	239	94	5.4	2
Post intubation	1	99.9	487	94	5.9	1.6
CPB	0.7	99.9	526	-	-	-0.9
post CPB	1	99.7	385	94	5.7	-1.8

Patient was shifted to ICU, where he was ventilated for 36 hours in 4 days ICU stay. With gradual reduction of Inj Dobutamine and he was shifted to ward on 5th postoperative day.

During ICU stay, Hb Electrophoresis was done to rule out Hemoglobinopathies and Photospectromery to rule out abnormal Hemoglobin compounds such as Meth Hb, Sulfa Hb or Hb M. Reports showed that patient had high levels of Meth Hemoglobin, 18 % as compared to 1.5% as normal value.

DISCUSSION

Dark color of arterial blood is cyanosis. Apart from Cardio Respiratory causes of Cyanosis, it can be because of abnormal Hemoglobin. The main causes remain as Carboxy Hb, MethHb, Sulf Hb and rarely Hb M.1,2

The difference between PaO2% on blood gas analysis and SpO2 % on pulse Oximetry, more than 5% should raise suspicion of abnormal hemoglobin. Some authors refer this as a "Saturation Gap".7

The diagnosis of Methhemoglobin is suggested by cyanosis in the presence of normal PaO2 but low measured SpO2. SpO2 & PaO2 discrepancy is there: so calculations using SpO2 for PaO2 which is normally matching may go wrong in presence of MethHb. Decreased SpO2 with normal PaO2 may alert Anaesthesiologist for presence of Meth Hb. Pulse Oximetry reading 85% regardless of the PaO2 is because of absorbance characteristic of Meth Hb.4

The routinely used pulse oximetry use a wavelength of 940mm for oxyhemoglobin detection and 660mm for Deoxyhemoglobin detection.

At low levels of Meth Hb (< 20%) Meth Hb is detected primarily by DeoxyHb sensor and at high levels (>70%) Meth Hb is primarily detected by Oxy Hb sensor.7

Standard Blood gas analyzers don't detect Meth Hb. They calculate oxygen saturation values from PaO2 and PH. To diagnose Meth Hb, Multiple Wavelength Co-Oximetry is used. As contrast to Pulse oximetry this is an invasive monitoring, requiring blood sample.7

For Cyanosis to be evident clinically, at least 5gm% of reduced Hb is required but it will be evident with 1.5% of Meth Hb and 0.5% of Sulfa Hb.1,2

All these forms reduce the capacity of hemoglobin to bind and carry O2.

CO Hb produces cherry bright color of blood and the levels of CO Hb will reduce with oxygen therapy in hours. In contrast MethHb and Sulf Hb produce Chocolate Brown and Mauve lavender color of blood respectively; don't respond to oxygen therapy.(1) But changes in color of blood can be differentiated only when high % present, otherwise Spectrophotometry can detect low conc. Of these compounds.2

Meth Hemoglobin is a hemoglobin in which Fe atom of Hemoglobin fails to remain in reduced state i.e. Fe+++ rather than normal Fe++ state. This causes inability to carry O2 by Hb. & also being is associated with leftward shift of Oxygen dissociation curve, O2 delivery to tissue is also decreased. Fe++ sate is maintained by ascorbic acid, Glutathine,, Methylene blue. Normal Meth Hb levels are less than 1.7%.

Meth Hb can be caused by a variety of chemicals, drugs and few congenital forms.

Industrial hazards from Aniline dye, Aminobenzines, Nitrobenzenes, Acetanilid can cause Meth Hb formation. Nitrous gas from arc welding, Nitrites from well water and food, Red wax from Crayons, Dyes from Blankets, Laundry marks, Shoes also can form Meth Hb.

Drugs like Sufonamides, Nitrites, Nitrates, Quinones are

important causes.

Congenital forms are DPNH Diaphorase Deficiency, TPNH Meth Hb Reductase deficiency, Decrease Glutathine formation in RBC's, Hemoglobin M. Congenital forms are rare.

Clinically to diagnose Meth Hb is difficult. Cyanosis occur only when Meth Hb levels reach 25% of total pigment. Anoxic symptoms occur at 30 to 40% level. Including Tachypnea, dizziness. Chronic Meth Hb can cause Polycythemia.

Electrophoresis differentiate Hb M by absorption wavelength which is shifted down than normal.2,

Photospectrometry diagnose and differentiate Meth Hb and Sulf Hb.

Sulf Hb is a rare cause of cyanosis & is due to abnormal sulfonation of Hb molecule. As contrast to Meth Hb which can be reverted to Normal form by Methylene Blue 1% injection, Sulf Hb can't be reverted to normal form. It gets terminated with RBC destruction only. Short or long term use of Metoclopramide is the important cause of Sulf Hb formation although drugs causing Meth Hb also can cause formation of Sulf Hb. As contrast to Meth Hb, the oxygen dissociation curve is shifted to right with sulf Hb so higher concentrations of Sulf Hb are well tolerated.

Abnormal hemoglobin, Meth Hb & Sulf Hb may be associated with altered spectra resulting in erroneously low SPO2 reading on pulse oximetry but Carboxy Hb is read by pulse oximetry as OxyHb sp readings may be normal or higher also.

In our patient with detection of Saturation gap, abnormal hemoglobin was suspected. But the cyanosis not reduced with prolonged oxygen therapy ruled out carboxy Hb. With suspicion of either meth Hb or Sulf Hb, the case was managed as per routine protocol: without any active intervention because:

1.SaO2 levels were normal. The absence of acidosis could give a clue of adequate oxygen delivery at tissue level.

2. The particular management of Sulf Hb is not known.1,3

3. Meth Hb should be treated if Meth Hb levels are high enough to impaired oxygenation which cause symptoms of oxygen deprivation, usually >30% Meth Hb levels.7,8
4. Before treating with Methylene Blue, G6PD deficiency should be ruled out.5

In this patient the use of Nitroglycerin was avoided. Also the obvious cause of Methemoglobinemia could not be found out from the patient history & investigations.

This is an effort to remind anaesthesiologist, "Dark colored arterial blood with low Spo2, not responding to increase in Fio2" can be an abnormal hemoglobin.

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Methemoglobinemia due to nitric oxide therapy in a child after cardiac surgery.

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