Sir MO, a 2 year old, previously healthy boy, presented to the emergency department at 2000 hrs with generalized cyanosis and agitation. A few hours prior to admission, his mother felt he looked pale and was acting strangely agitated. Later, however, he fell asleep. When he woke up, his mother noticed he was deep blue in colour. He vomited on his way to the hospital. On arrival to the emergency room he was restless, deeply cyanosed and incoherent.

He was started on high flow oxygen via non-rebreather. Pulse oximetry showed oxygen saturation of only 71%.

**Vital Signs**
- Temperature: 37°C, HR: 151/min, RR: 26/min, BP: 159/79mmHg, repeat 136/71mmHg.
- Head and neck examination was unremarkable. Chest was clear to auscultation. There was no respiratory distress noted. Cardiovascular examination revealed normal heart sounds with no murmurs audible. Abdomen was soft, non-tender with no hepatosplenomegaly.

**Initial investigations revealed**
- FBC: Hb. 101 g/dl, WCC 11.0 x10⁹/L, PLT 326 x10⁹/L.
- Na: 138mmol/L, K: 3.5mmol/L, Cl: 104mmol/L, Urea: 2.45mmol/L, Alb: 46g/L, Phosphate: 1.25mmol/L, Venous blood gas: pH 7.33, pCO₂ 5.99kPa, pO₂ 2.18kPa, HCO₃ 21.2mmol/L, BE 1.7mmol/L.

Intravenous access was established and oxygen continued. When asked repeatedly about any possibility of accidental ingestion, MO's mother revealed that his grandmother who was visiting then was on dapsone tablets for dermatitis herpetiformis. She made contact with her mother who said there was 5-7 tablets missing from her medicine container and the lid of her container was open.

A diagnosis of methaemoglobinaemia was made secondary to ingestion of dapsone.

MOs serum methaemoglobin level was reported at 52%, (normal range 1-2%). He was given IV methylene blue (1mg/kg) with gradual resolution of cyanosis, worsening cyanosis with raised levels of methaemoglobinaemia, increasing agitation, and hallucinations. He improved significantly over 48 hours and was then transferred to the ward.

**Discussion**
Dapsone is a sulfone active against a wide range of bacteria but mainly employed for its action against mycobacterium leprae. It inhibits folic acid in susceptible organisms. It is used with pyrimethamine in the treatment of malaria. Dapsone is also used in the treatment of dermatitis herpetiformis, occasionally for pyoderma gangrenosum, pustular psoriasis and acne vulgaris. Dapsone overdose can result in severe and prolonged clinical effects through interaction with both haemoglobin and red cells resulting in methaemoglobinaemia and later haemolysis. Peak blood levels occur 2-6 hours post ingestion and it has a prolonged half life, as it is highly protein bound and lipid soluble. Onset is within a few minutes to 24 hours. Methaemoglobinaemia is a major clinical effect with cyanosis of the mucus membranes, nails and extremities. This may last 3-10 days and is unresponsive to oxygen therapy. Other symptoms can include dizziness, confusion, agitation, hallucinations, and delirium.

Methaemoglobin levels should be carefully monitored. A level greater than 20% requires treatment. Methylene blue (tetrathiomethylthionine chloride) should be administered in doses of 1-2 mg/kg as a slow bolus over 5 minutes. Total doses of methylene blue should not exceed 7mg/kg, as this itself can cause methaemoglobinaemia.

The administration of methylene blue would be contraindicated if the patient has underlying G-6-P-D deficiency as it can induce haemolysis.

The degree of clinical cyanosis should not guide therapy as methylene blue itself can cause grayish blue discoloration of the skin.

This case illustrates the importance of considering the diagnosis of methaemoglobinaemia in a child presenting with generalized cyanosis, especially when the presentation is acute in a previously healthy child.

**Correspondence:**
Farhana Sharif,
17 Boyne View, Johnstown,
Navan, Co Meath, Ireland.
Email: farhanasharif@eircom.net

**References**