



## Left lower Limb Gangrene Following Diarrhoeal Disease and Dehydration in a child with Transposition of the Great Arteries

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### Abstract

**Background:** Children with Cyanotic Congenital Heart Diseases are at risk of developing thrombosis and gangrene, especially when they suffer severe fluid loss or experience significant fluid restriction.

**Aim:** To describe a case of a 2-year-old girl with transposition of the great arteries (TGA) who developed left lower limb gangrene following an episode of diarrhoeal disease.

**Case:** HB presented with four-week history of progressive darkening of her left leg, which was preceded by two days history of diarrhea and vomiting. At presentation, she had moderate dehydration and central cyanosis (SpO<sub>2</sub>-68%), with signs of dry gangrene of the distal half of the left leg. Echocardiography confirmed d-TGA with ventricular and atrial septal defects. Packed cell volume was moderately elevated (57%), but blood culture result and other hematologic parameters were essentially normal. Patient had above-knee amputation of the left leg. She had eventful post-operative stay characterized by inter current fever and delayed wound healing (>4weeks). Unfortunately, the parents signed against medical advice despite counseling.

**Conclusion:** Limb gangrene may complicate cyanotic congenital heart diseases such as TGA. Diarrhoeal disease, which causes dehydration and hyper viscosity, is a potential precipitant.

**Keywords:** Gangrene, Transposition of Great Arteries, Diarrhoeal Disease, Dehydration, Sokoto, Nigeria.

### Introduction

Transposition of the Great Arteries (TGA) is a Cyanotic Congenital Heart Disease (CHD) in which the pulmonary artery arises inappropriately from the left ventricle and the aorta arises from the right ventricle, leading to systemic oxygen desaturation and tissue hypoxia.<sup>1</sup> Like any other cyanotic CHD, affected children are prone to thrombo-embolic events, which may lead to ischaemic necrosis and gangrene.<sup>1,2</sup>

Polycythaemia is a relatively common complication in cyanotic patients with CHD and it can lead to thrombosis especially when associated with iron deficiency, sepsis or significant dehydration.<sup>1</sup> Limb gangrene following dehydration is uncommon. Only a few cases have been reported in the literature.<sup>3,4</sup> Morin and Chevalier had reported a case of right lower limb gangrene necessitating amputation in a neonate with hypernatraemic dehydration.<sup>4</sup>

In Nigeria, available reports of extremity gangrene were mainly in children with normal heart; and the cause for this was attributed to many factors such as sepsis, disseminated intravascular coagulopathy (DIC) and intrauterine thromboembolic event.<sup>5-7</sup> Though Animasahun and Amoah<sup>8</sup> had reported a case of upper limb gangrene in a Nigerian child with tetralogy of Fallot, the precipitating factor was presumably an embolus from endocardial vegetation rather than dehydration. We present a case of Transposition of the Great Arteries (TGA) in a 2-year old girl, who developed left lower limb gangrene following an episode of diarrhoeal disease with dehydration. The aim is to emphasize the risk of dehydration, an easily preventable problem, to a child with cyanotic CHD.

### Case

HB is a 2-year-old girl who presented with four-week history of progressive darkening of her left leg. The symptom was noticed two days after the onset of profuse watery diarrhoea and vomiting. She was initially treated at a primary health center (PHC) with Oral rehydration solution (ORS), metronidazole and other medications. Diarrhoea and vomiting were said to have subsided after a week; but the limb discoloration gradually worsened, which warranted referral to our hospital. On general examination at presentation, she was afebrile, small for age and moderately dehydrated. She has conjunctival injection, grade 3 digital clubbing and central cyanosis. Her oxygen saturation was 68%. Cardiovascular examination revealed mild precordial bulge, with the apex beat located at 5<sup>th</sup> left intercostal space lateral to mid-clavicular line. She has grade 3/6 systolic murmur. There was darkish discoloration of the distal half of her left leg extending to the left foot, in keeping with dry gangrene as shown in figure 1.

Investigations included Echocardiography, which confirmed transposition of the great arteries (TGA) with associated ventricular and atrial septal defects. Packed cell volume was 57%. Full blood count and differentials were within normal limit

while blood culture yielded no growth. After counselling of the parents, above-knee amputation of the left leg was performed. She however had eventful post-operative stay, with repeated episodes of worsening oxygen desaturation, intercurrent infection and significant delay in wound healing (>4weeks). The parents signed against medical advice despite repeated counselling.



**Figure 1:** Gangrene of the distal half of the left leg in a 2 year old girl with TGA.

### Discussion

This report highlights an unusual case of peripheral gangrene following diarrheal disease in a child with Transposition of the Great Arteries. There are many factors that could potentially lead to development of thrombosis and even gangrene in children with cyanotic CHD.<sup>2,9</sup> These include polycythemia, infective endocarditis, central embolism, sepsis, dehydration and DIC which may either be induced by infection or immune-mediated reactions.<sup>2,8</sup>

In our patient, dehydration was presumed to be the most likely precipitating factor in view of the temporal relation between the onset of diarrhoea and the occurrence of limb gangrene. Severe dehydration either due to significant fluid loss or fluid deprivation has been identified as one of the risk factors for gangrene in children.<sup>3,10,11</sup> One of the earliest documented evidence that suggested such association in literature was an old report by Comayet *al.*<sup>10</sup> Similarly, Bass and colleagues<sup>11</sup>

had reported cases of extremity gangrene complicating diarrheal disease and dehydration. In neonates and young infants on exclusive breastfeeding, occurrence of peripheral gangrene was also linked to hypernatraemic dehydration resulting from inadequate fluid intake.<sup>4,12</sup>

As explained by Kotby *et al*<sup>2</sup>, dehydration causes hypovolaemia and haemoconcentration, which can precipitate hyperviscosity. The latter results in stasis that may lead to hypercoagulable state and tendency for thrombus formation.<sup>2</sup> Hence, it is necessary to ensure optimum hydration of all children with diarrhoea or other conditions associated with fluid loss, particularly if they have underlying cyanotic congenital heart diseases.

Polycythemia is a common complication in patients with cyanotic CHD, and occurs as a physiologic response to tissue hypoxia, which induces increased bone marrow erythropoiesis.<sup>1,9</sup>

Polycythemia can precipitate thrombosis especially when there is associated hyper viscosity due to dehydration or iron deficiency; or in the presence of other risk factors such as sepsis and DIC.<sup>1,5,8</sup> Though our patient's hematocrit level was only moderately elevated, she still developed peripheral gangrene most likely because of the associated dehydration, which served as an additional precipitating risk factor.

It is of note that the role of elevated haematocrit as a sole risk factor for thrombosis in patients with cyanotic CHD or secondary polycythemia has recently been questioned.<sup>9,13,14</sup> Unlike in patients with polycythemia vera, current literature showed no direct correlation between increased hematocrit level and risk for thrombogenesis.<sup>14</sup> Other workers have reported peripheral gangrene resulting due to other causes such as septicaemia and DIC.<sup>5-8, 15, 16</sup>

These causes were however not evident in our patient. Unlike Animasahun and Amoah<sup>8</sup>, who reported infective endocarditis (IE) leading to upper limb gangrene in a child with TOF, there was no indication of IE in our patient from the results of echocardiography assessment and serial blood culture analysis. We were unable to undertake detailed hematological assessment to

exclude DIC in our patient. Nevertheless, available clinical and laboratory evidence does not seem to support this possibility in the patient.

### Conclusion

Diarrheal disease with dehydration can cause peripheral gangrene in children with cyanotic congenital heart disease. Early diagnosis and prompt correction of dehydration would go a long way in preventing this pathetic, but avoidable complication.

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