Nephrotic Syndrome in a Child Suffering from Tetralogy of Fallot: A Rare Association

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Nephrotic syndrome is an uncommon complication of tetralogy of Fallot and has been rarely reported in pediatric population. We describe a 4-year-old female Congolese child who was referred for investigation for persistent dyspnea, edema, and cyanosis and nephrotic range proteinuria. Our patient presented with a tetralogy of Fallot and nephrotic syndrome. Conclusion. This case reminds us that children with tetralogy of Fallot may develop nephrotic proteinuria.

1. Introduction

Glomerular dysfunction can be found in cyanotic congenital heart disease (CHD) especially in older children and adults, being associated occasionally with proteinuria and microalbuminuria [1, 2]. The risk of developing renal impairment is particularly high in cyanotic patients particularly in patients with long-standing cyanotic CHD [1, 2]. However, nephrotic syndrome (NS) is an uncommon complication of cyanotic congenital heart disease and is rarely reported. This complication has not been documented in Congolese children.

2. Case Report

A 4-year-old girl with edema, dyspnea, and cyanosis was referred from Butembo at the East of the Democratic Republic of Congo (DRC) to a facility renal and cardiology investigations in Department of Pediatrics of University Hospital of Kinshasa, Kinshasa, DRC. The history of present illness dates back to about 13 months characterized by progressive cough, dyspnea, and orthopnea. Physical examination revealed respiratory distress with edema and episodes of squatting. She was cyanosed with finger and toes clubbing. Apex beat was at the fifth intercostal space anterior axillary line. Both heart sounds were noted with a systolic thrill and loud systolic ejection murmur grade 3. The blood pressure was 150/110 mmHg, and oximetry was 63%.

A complete blood count showed hemoglobin 23.2 g/dL, total proteins 45 g/L, and albumin 22 g/L. Dipstick urinalysis was 3+ while the 24-hour urinary protein was 154 mg/kg. Creatinine was 37 μmol/L, urea was 3.8 mmol/L, cholesterol was 4.3 mmol/L, and HDL was 1.3 mmol/L. HIV and hepatitis serology were negative. Anti-streptolysin O (ASLO) was <200 IU.

X-ray revealed “boot shaped” heart with an upturned cardiac apex (Figure 1). Echocardiography revealed tetralogy of Fallot with hypoplastic pulmonary artery and biventricular dysfunction. Cardiac catheterization was not performed due to technical reasons. A diagnosis of Fallot's tetralogy and NS was established. Renal biopsy was contraindicated because of the deteriorating renal condition and cardiac status.

During her hospitalization, the child received specific treatment for her blood hypertension, associated furosemide (1 mg/kg/dose, every six hours), Propranolol (2 mg/kg, every
Table 1: Results of the literature review of cyanotic CHD associated with nephrotic syndrome in African children.

<table>
<thead>
<tr>
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<tr>
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<td>12</td>
<td>7</td>
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<tr>
<td>Gender</td>
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<td>Male</td>
<td>Female</td>
<td>Male</td>
</tr>
<tr>
<td>Cyanotic CHD*</td>
<td>Tetralogy of Fallot</td>
<td>Truncus arteriosus</td>
<td>Tetralogy of Fallot</td>
<td>Tricuspid atresia</td>
</tr>
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*CHD: congenital heart disease; DRC: Democratic Republic of Congo.

Figure 1: Plain film shows a "boot shaped" heart with an upturned cardiac apex due to right ventricular hypertrophy and concave pulmonary arterial segment.

The patient presented late. This case revealed the problem of early diagnosis and management of cyanotic CHD in resource-limited settings as in DRC. Considering the paucity of facilities available for medical and surgical management of Fallot's tetralogy in our midst, we recommend early detection of this congenital heart disease and regular renal screening of patients and thus allow, at least at this stage, the initiation of ACE-I. However, it has to be stated that, in countries such as the DRC, early corrective cardiac surgery should be the first choice particularly in

Mechanisms have been suggested for the development of nephropathy. The patients with cyanotic CHD are exposed to chronic hypoxia. The risk of developing glomerular lesions rose sharply during the second decade of life if the cyanosis remains unchanged for more than ten years [4, 5]. Hyperviscosity due to polycythemia may induce an angiogenic increase in the glomerular capillary beds, in turn leading to glomerulomegaly. Glomerulomegaly is a consequence of the hyperperfusion of glomeruli associated with the chronic hypoxia and the increased hydrostatic pressure in the capillary wall. This situation is a causative factor of the deterioration and the decline of renal function, in the condition of polycythemia. Furthermore, the failure of a compensatory mechanism to respond to reduced RPF by hyperfiltration may be accompanied by the development and progression of microalbuminuria and proteinuria [4–6]. The pathogenesis and development of nephrotic proteinuria range are the result of these combined mechanisms. Although we could not perform the renal biopsy, the nephrotic range proteinuria is probably a consequence of focal and segmental glomerulosclerosis.
patients who otherwise present severe complications of their cyanotic CHD and reduce the risk of the development of chronic renal failure.

**Ethical Approval**

This study was determined to be Non-Human/Non-Research by the Ethical Committee of the Public Health School of the University of Kinshasa, Kinshasa, DRC.

**Conflict of Interests**

The authors declare that there is no conflict of interests regarding the publication of this paper.

**References**


