

## CASE REPORT

# Intracardiac thrombus – a rare complication of the steroid resistant nephrotic syndrome

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**Abstract:** *Background:* An intracardiac thrombus is extremely rare in children with the nephrotic syndrome (NS).

*Objectives:* To present a case report of a child with steroid resistant NS and intracardiac thrombus.

*Methods and results:* A 3.5 year-old boy with the first attack of steroid resistant NS was admitted to the hospital. A histological evaluation of the renal biopsy specimen revealed the minimal changes disease (MCD). There were no mutations in the podocin gene. The treatment with furosemide, albumin, prednisone, methylprednisolone, cyclophosphamide, enalapril and losartan was ineffective, as the intermittent oedema, proteinuria, hypoalbuminemia and hypercholesterolemia were still present. 8 weeks after the disease onset, the sinus tachycardia occurred and the echocardiography revealed a thrombus in the right ventricle, which had gradually proceeded to the pulmonary artery bifurcation. The thrombolysis with 40 mg of alteplase was initiated. Two hours after the alteplase application, the thrombus was not detectable. The mutational analysis of factors V, II and MTHFR genes were negative. The repeated echocardiography performed after 1 week, 2 and 6 months, respectively, revealed a normal cardiac function and morphology. The patient received prophylactic doses of fraxiparin for 3.5 months followed by warfarin. A remission of the nephrotic syndrome was achieved with high doses of cyclosporine A together with atorvastatin at 7 month after the disease onset.

*Conclusion:* The thromboembolism as a result of the hypercoagulation status is a serious complication of the nephrotic syndrome. The intracardiac localisation of thrombus is extremely rare (Fig. 2, Ref. 10). Full Text (Free, PDF) [www.bmj.sk](http://www.bmj.sk).

Key words: intracardiac thrombus; steroid-resistant nephrotic syndrome.

Thromboembolic complications (TEC) are rare and the most serious complications in children with the nephrotic syndrome (NS) (1–4). We present a case of the steroid resistant nephrotic syndrome, where an intracardiac thrombus was detected in the right ventricle.

## Case report

A 3.5 year-old boy with the first attack of the steroid resistant NS was admitted to the hospital in May 2005. At admission, there was a normal echocardiographic finding with the bilateral pleural effusion and ascites on the abdominal ultrasonography. A histological evaluation of the renal biopsy specimen revealed the minimal changes disease. There were no mutations in the podocin gene. The treatment with furosemide, albumin, prednisone, methylprednisolone, cyclophosphamide, enalapril and

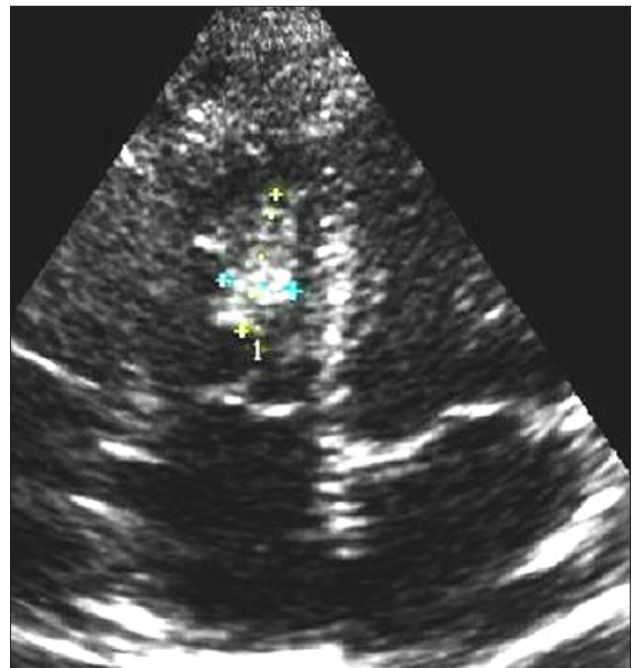


Fig. 1a. Echocardiographic image before thrombolytic treatment – thrombus in the right ventricle.

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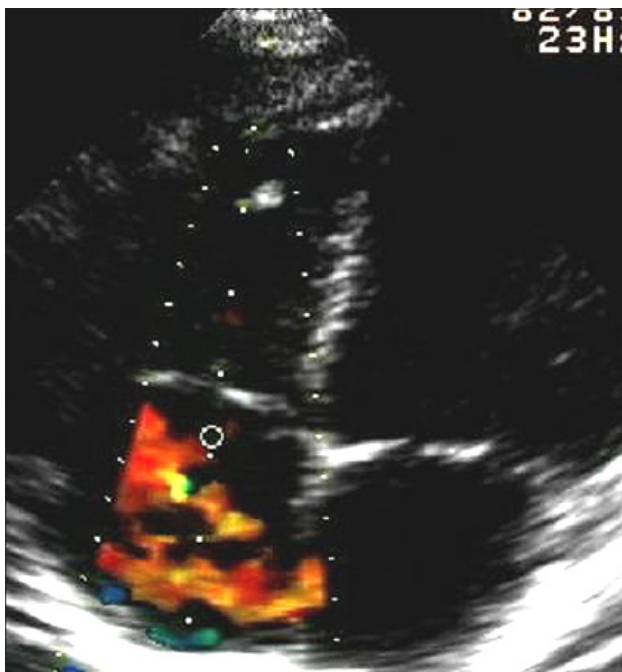


Fig. 1b. Echocardiographic image after treatment.

losartan was ineffective, as the intermittent oedema, proteinuria (31.6 g/24 h), hypoalbuminemia (15.6 g/L) and hypercholesterolemia (20.5 mmol/L) were still present. 8 weeks after the disease onset, the sinus tachycardia occurred (140 beats/min). The echocardiography revealed a primary thrombus (20x11 mm) in the right ventricle, reaching the right ventricular outflow tract, moderately influencing ventricle function. The patient was transferred to the Cardiocentrum of the University Hospital Motol in Prague. At that time, the thrombus has been already detected in the pulmonary artery bifurcation (Fig. 1a). Therefore, the thrombolysis with 40 mg of alteplase, which is a tissue-type plasminogen activator, was started. Two hours after the alteplase application, the thrombus was not detectable. The lung perfusion scan performed 5 days after the therapy initiation revealed a normal appearance of the right lung and a large multisegmental perfusion defect of the left lung, affecting mostly the lower lobe and a part of the lower segment of the upper lobe (Fig. 2). Haematologic parameters were following: thrombocytes  $474 \times 10^9/L$ , fibrinogen 12.05 g/l (normal 2–4 g/l), antithrombin III 28 % (n 80–120 %), protein C 213 % (n 70–140 %), protein S 100 % (n 65–140 %), INR index 0.97, APTT index 1.4, S-homocystein 12.5  $\mu\text{mol/l}$  (n 9.2–15.0  $\mu\text{mol/l}$ ). The mutational analysis of factors V, II and Methylene-Tetra-Hydro-Folate-Reductase (MFTHR) genes were negative. The repeated echocardiography performed after 1 week, 2 and 6 months, respectively (Fig. 1b), revealed a normal cardiac function and morphology. The patient received prophylactic doses of fraxiparin for 3.5 months and antithrombin III, followed by 4 months of warfarin. A remission of the nephrotic syndrome was achieved with high doses of cyclosporine A 10 mg/kg/day together with atorvastatin 10 mg/day at 7 month after the disease onset.

## Discussion

TEC is a serious complication of NS, however it may go unnoticed and asymptomatic or with a subclinical course. The hypercoagulation status in NS might occur due to antithrombin III (ATIII) deficiency, increased fibrinogen level, decreased protein C and S activity, increased platelet number and aggregation. The risk factors for TEC in patients with NS are hypovolaemia, diuretic therapy, infection, immobilization, trauma, corticosteroids, hypoalbuminaemia  $<20 \text{ g/L}$  and proteinuria  $>10 \text{ g/L}$  (1, 2). The reported incidence of TEC is relatively high in the nephrotic adults, ranging from 9 % to 70 %, and often occurring in patients with the membranous glomerulonephritis. The incidence of clinical vascular TEC in children with NS is reported to be 1.8–5.3 %, but it may be higher (1.5–66 %) when these specific complications are searched for. Paediatric patients with the secondary forms of NS have a higher incidence of thrombotic events than those with the minimal change disease (1). In a large retrospective study of 447 children with NS, the incidence of TEC was 2 % (9/447); 16 clinically apparent TEC were registered in 9 children. The reported incidence of TEC was 1.5 % among patients with the steroid-sensitive NS and 3.8 % among those with the steroid-resistant NS. TEC were usually venous – 81 % versus 19 % located in the arteries (2). The intracardiac location of thrombus is extremely rare (3–10), as thrombi form mostly in deep leg veins, inferior vena cava, and rarely in superior vena cava, renal veins, axillary, subclavian, femoral, coronary and mesenteric arteries, and hepatic veins (1–4). So far, the intracardiac thrombi have been reported in eight patients (3, 5–10) – in three adults (5–7) and five children (3, 8–10). In our patient, the TEC occurred in the primary steroid-resistant NS. The relatively

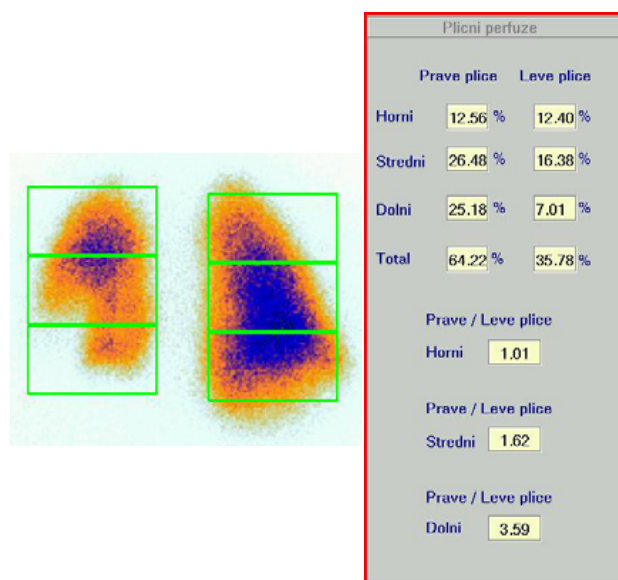


Fig. 2. Lung perfusion scan performed 5 days after the initiation of thrombolytic therapy. Normal appearance of the right lung and large multisegmental perfusion defect of the left lung. Right to left lung perfusion ratio 64.22 % : 35.78 %.

early detection of the intracardiac TEC in our patient led to the rapid onset of thrombolytic therapy and dissolution of the thrombus with no serious consequences. Paediatric nephrologists should be alerted on the possibility of TEC in various locations.

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