Spontaneous resolution of congenital nephrotic syndrome in a neonate

C R Banton, B Thalayasingam, M G Coulthard

Abstract
An infant with congenital nephrotic syndrome recovered spontaneously and completely by the age of 11 days and had remained well at the age of 1 year. This reinforces the view that reversible congenital nephrotic syndrome does occur and that it is not a single disease with a universally dismal prognosis.

Congenital nephrotic syndrome presents in the first three months of life,1 2 and usually results in death in early childhood unless intensive treatment including renal transplantation is undertaken.1 3

Only four cases of congenital nephrotic syndrome with spontaneous recovery have previously been reported, and of these two did so at 10 and 25 months.1 3 The other two were siblings whose mother had focal segmental glomerulosclerosis and were presumed to have been affected by humoral factors; they recovered in less than three weeks.4

---

Spontaneous resolution of congenital nephrotic syndrome in a neonate

Case report
A girl presented at the age of 92 hours with peripheral oedema. She was the first child of a 29 year old white mother; the pregnancy had been uncomplicated, with normal blood pressure, no proteinuria, and she had taken no medication. Family history was unremarkable. She was born at 38 weeks' gestation weighing 2466 g, and the 1 minute Apgar score was 9. The placenta weighed 640 g and was healthy.

Examination at 12 and 48 hours (because of poor feeding) showed no abnormality. By 92 hours she had only lost 16 g despite feeding poorly and she had developed oedema of the limbs and around the eyes. Her pulse was 160/min, respiratory rate 40/min, and blood pressure 75/49 mm Hg.

The urine looked clear, but there was gross haematuria and proteinuria on urinalysis. There were no casts present. Urinary electrophoresis showed a heavy unselective proteinuria. The plasma protein concentrations were low; total protein was 30 g/l (albumin 20 g/l and globulin 10 g/l), confirming the diagnosis of nephrotic syndrome. Investigations did not reveal a cause for the congenital nephrotic syndrome; antenatal Veneral Disease Research Laboratory screening for syphilis was negative as were the baby’s toxoplasma, rubella, herpes and cytomegalovirus titres. She had a normal female karyotype. Ultrasound scanning showed slight bilateral renal enlargement (kidney length 5-5 cm) with poor corticomedullary differentiation and no evidence of hydronephrosis or renal vein thrombosis. Her mother’s urinary albumin:creatinine concentration ratio was normal at 1-9 mg/mmol (upper limit of reference range 3-5).

Over the next two days the baby’s feeding improved, but she developed increasing oedema, gained 500 g in weight, and showed clinical signs of intravascular hypovolaemia with strikingly cold peripheries. Biochemical and haematological indices were consistent with hypovolaemia. Her plasma creatinine concentration was normal at 38 μmol/l, and the sodium was low at 126 mmol/l. Her urinary sodium was less than 5 mmol/l, and the urine: plasma ratio for urea was greater than 12 giving a value for fractional reabsorption of sodium of at least 99-6%. The haemoglobin concentration was 190 g/l.

She was treated with 15 ml/kg of plasma protein fraction and 2 mg frusemide given intravenously, which resulted in improved peripheral perfusion, a diuresis, and loss of 170 g in weight, a fall in haemoglobin concentration to 170 g/l, and correction of her plasma sodium concentration to 144 mmol/l.

Her proteinuria remained (maximal colour on test strip) for three days, then reduced gradually; by day 11 her urine contained no protein, her plasma albumin concentration had risen to 31 g/l, her oedema had completely resolved, and she was feeding well and gaining weight. A repeat ultrasound scan at 19 days showed kidneys of normal length (4-4 cm) with corticomedullary differentiation. At 1 month a dimercaptosuccinic acid scan was normal. At 1 year she remained completely well, with a urinary microalbumin concentration below the detection limit of the assay (12 mg/l).

Discussion
As this baby had proteinuria, hypoproteinaemia, and oedema at 4 days of age, she had by definition got congenital nephrotic syndrome. Renal biopsy was not carried out because of her spontaneous recovery, and no predisposing cause was identified. Only four cases have previously been described in which spontaneous resolution of congenital nephrotic syndrome occurred; in two this was after much longer (10 and 25 months),1 3 and in one of these hypotensive treatment was required initially.7 The other two cases were siblings whose mother had focal segmental glomerulosclerosis; the index case’s mother had normal urinary protein excretion. All other cases of congenital nephrotic syndrome that have resolved have required treatment with steroids,2 or have been infants with congenital syphilis who have been treated with penicillin.

Congenital nephrotic syndrome is associated with various histological patterns, of which the Finnish type is most common. Others include diffuse mesangial sclerosis, focal segmental glomerulosclerosis, and—rarely—a pattern of minimal change seen in infants that have responded to treatment with steroids.8 As congenital nephrotic syndrome is usually an irreversible debilitating condition that leads to death unless active treatment is instituted, it is likely that these rare cases that undergo spontaneous remission represent a separate histological subgroup. To our knowledge this girl had the earliest spontaneous recovery yet described. Her case reinforces the view that reversible congenital nephrotic syndrome does occur, and that congenital nephrotic syndrome is not a single disease with a universally dismal prognosis.

Spontaneous resolution of congenital nephrotic syndrome in a neonate.

C R Banton, B Thalayasingam and M G Coulthard

*Arch Dis Child* 1990 65: 992-993
doi: 10.1136/adc.65.9.992