Urethral Prolapse in Girls

The purpose of this short report is to draw attention to a condition known to gynaecologists and urologists, but apparently unfamiliar to many paediatricians.

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Case 1. A 6-year-old girl had noticed painless bleeding from the vulva for three weeks, unassociated with micturition. Examination showed a well-developed West Indian girl with a reddish purple swelling at the urethral orifice. Examination under anaesthesia revealed oedema and inflammation of prolapsed urethral mucosa with some discharge, from which Staphylococcus pyogenes was cultured. Catheterization of the bladder produced turbid urine which was, however, uninfected. Hb 9.9 g/100 ml; other investigations normal. She was treated with a course of oral ampicillin and local applications of hydrocortisone and neomycin for one month. There has been no evidence of relapse after two years.

Case 2. For two weeks a vulval mass had been noted in this 6-year-old girl. This was painless but she had had intermittent haematuria for the same period. Examination revealed a pink prolapse of the urethra. Bladder catheterization produced clear, uninfected urine, and biopsy revealed thickened epithelium infiltrated with polymorphs. The prolapse was excised. A year has elapsed without recurrence of symptoms.

Case 3. Blood stains on the underclothes were noted a day before and again on the morning of admission in this 5-year-old girl. Past health had been good apart from recurrent mild epistaxis. Examination revealed a reddish purple mass at the urethral orifice, which bled on being touched. Hb 12.6 g/100 ml, WBC 4500/mm³ with a normal differential count; platelets 248,000/mm³. Bleeding time, clotting time, and prothrombin index normal. Examination under anaesthesia confirmed the diagnosis of urethral prolapse, and the bladder was catheterized with the production of clear, uninfected urine. Hydrocortisone and neomycin was applied locally and the child was discharged in 48 hours, much improved. Local treatment was continued for a month, and two months later there was been no sign of relapse.

Discussion

Though urethral prolapse was first described nearly 250 years ago, probably less than 400 cases have been recorded. Säuerlin (1929) collected all the examples he could find up to that date and reported on 270. He analysed 211 patients with the condition, almost exactly half of whom were children. Since that time there have been a number of single case reports but only one large series, that of Owens and Morse (1968) which dealt with 54 children. The early authors recognized that the condition occurred either in childhood or old age, and rarely between puberty and 60 years. The youngest child reported was only 5 days old (Barnes, 1953). Recently American authors, Peters (1962) and Owens and Morse (1968), have noted a predilection for Negro girls, and this may have some bearing on the apparent infrequency of examples in this country in the past, and it is noteworthy that our three girls were West Indians. This racial preference does not seem to affect elderly patients.

The usual method of presentation is with bleeding, usually only enough to soil clothing, though occasionally haematuria may occur (as in Case 2). Sometimes the bleeding is mistaken for the onset of menstruation. Urinary symptoms are infrequent, but scalding micturition, frequency, incontinence, and retention have all been described. The children are not ill and their activity is unimpaired.

The diagnosis is made by inspection, when the prolapsed urethral mucosa will be seen as a non-tender mass surrounding a central urethral orifice. The colour varies from pale red to black, depending upon whether the mucosa is merely oedematous or has progressed to gangrene.

Histology of the lesion varies from simple oedema to dilated and thrombosed veins with or without superadded inflammation. Diagnosis is not difficult as long as one remembers that the prolapse is circular and encloses a central urethral orifice. This appearance excludes caruncle which only affects one segment of the urethra. A urethral polyp will be seen to protrude from the orifice. Sarcoma botryoides, ureteroceles, neoplasm, or condylomata might enter the differential diagnosis, but none of these shows the essential characteristic of a circular mass with a central orifice.

The reason why the condition is seen at the extremes of life and rarely during the reproductive age is unknown. Epstein and Strauss (1937) point out that a relatively long and perpendicular urethra
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held by loose connective tissue makes it mobile and, in children, the mucous membrane may have developed more strongly than the muscularis mucosae. They concluded that two factors might operate: first, a congenital weakness of tissue with an unduly wide urethra and, secondly, an acquired factor such as debility or inflammation. Other theories frequently mentioned are redundancy of the urethral mucosa, stress from cough or straining at stool, straddle injury, foreign body, or a true urethral hernia. Keefe (1917) favoured a neuromuscular cause, whereby the constrictor muscle is too weak to oppose a strong detrusor.

Many different methods of treatment have been used. Conservative treatment with replacement of the prolapsed mucosa, followed by continuous pressure, has been advocated but the tendency to relapse is high. A variation of this method is to insert a large catheter after replacing the mass. Catheter drainage is instituted for two to three weeks but relapses still occur after this procedure. Perhaps the most commonly used surgical treatment is to pass a catheter without making any attempt to reduce the mass. The redundant mucosa is then ligated at the base, over the catheter, and allowed to separate. Alternatively, it may be amputated with diathermy. Peters (1962) favoured fulguration at the ‘four points of the compass’ to induce submucosal fibrosis. Hepburn (1927) used an ingenious method whereby the prolapse was reduced by pulling it up the urethra by a suprapubic approach and stitching the bladder neck to the pubic bone. However, this method has only been considered for elderly patients, and the other surgical measures seem to have given good results with infrequent relapses. Conservative treatment with the local application of steroid and an antibiotic has shown promise in two of the cases, but the time is too short and the number too few to draw any conclusions.

Summary

Three examples of urethral prolapse in West Indian girls are described. This condition should be suspected when haematuria or vulval bleeding occurs in a West Indian girl.

References


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Chronic Copper Poisoning Presenting as Pink Disease

Copper toxicity is extremely rare, and Sternlieb and Scheinberg (1964) suggest that this is due to excretion or incomplete absorption of the metal rather than to an inherent lack of toxicity in copper. It is only in Kinnier-Wilson's disease (hepato-lenticular degeneration) that progressive copper toxicity is likely to occur due to defective copper balance from deficiency of copper-binding caeruloplasmin. Acute copper poisoning (usually copper sulphate in weed-killers) is not infrequently encountered but, as far as we are aware, no case of poisoning due to chronic ingestion of copper has so far been reported. With increased use of copper for hot-water pipes, further examples of copper poisoning must be anticipated.

Case Report

A 15-month-old infant was admitted to the Children's Unit after a 5-week history of behaviour change, diarrhoea, and progressive marasmus. Before admission developmental progress had been entirely satisfactory and there was nothing of significance in the birth or family history. When first seen the clinical picture was that of pink disease with prostration, misery, red extremities, hypotonia, photophobia, and peripheral oedema (Fig. 1). The body weight was only 7·48 kg (weight at 1 year 10·09 kg). The liver was palpable 2 cm below the costal margin.

Investigations. The outstanding finding was a raised serum copper level of 286 µg/100 ml (normal range taken as 164±70 µg/100 ml (Zak, 1958)), with a urinary copper level of zero (normal range 48±16-3 µg/100 ml (Eden and Green, 1940)). The serum and urinary copper levels were measured in a laboratory where these estimations are routinely performed, and where a high level of accuracy can be expected. All tests for mercury were negative. Other investigations which gave abnormal values were: serum aspartate aminotransferase (SGOT) and alanine aminotransferase (SGPT) both 180 Karmen units/100 ml. Alkaline phosphatase 33 K A units. Lactic dehydrogenase, γ-glutamyl transpeptidase, and pseudocholinesterase activity were not increased. Total plasma proteins were 4·0 g/100 ml with an albumin level of 2·2 g/100 ml.