A rare case of Pseudohypoaldosteronism in a neonate secondary to Congenital Hydrometrocolpos

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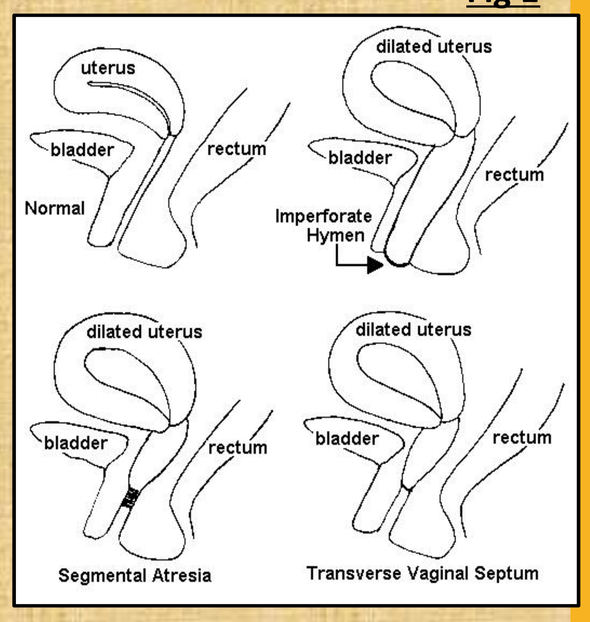
Introduction

- **Hydrometrocolpos;** There is distension of the vagina & uterus due to accumulation of secretions (other than blood), caused by increased oestrogenic stimulation & vaginal outflow obstruction.
- Incidence in term neonates is **0.00014-0.001%.**
- It presents **at extremes of childhood,** at birth when maternal circulating hormones are raised & at menarche when oestrogen production commences**.**
- **Pseudohypoaldosteronism (PHA)** due to obstructive uropathy, urinary tract infections & congenital urogenital malformations (fig 1) have been reported in the literature, however, there are no reports of PHA associated with hydrometrocolpos due to a common urogenital tract

Fig 1



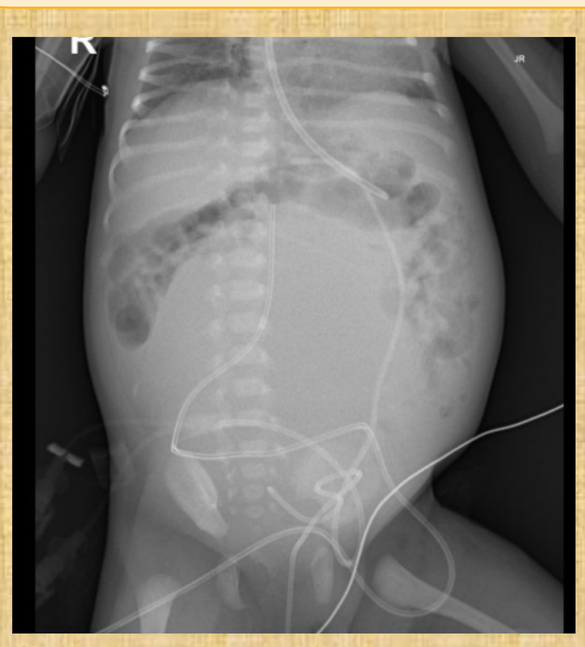


Fig 2

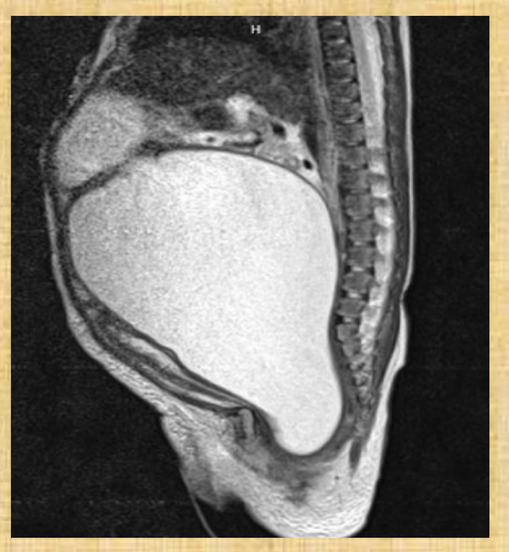
First Presentation

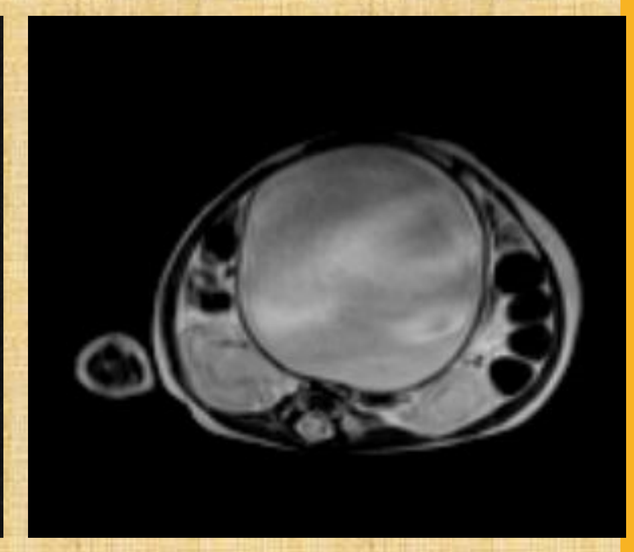
A female baby, 35 weeks gestation, birthweight of 2100g, was noted to have an abdominal mass soon after birth, with an abnormal x-ray (fig 2).

Ultrasound scans showed a round, central abdomino-pelvic cystic mass with bilateral renal pelvic-calyceal dilatation & hydronephrosis. An MRI (fig 3) showed huge distention of the vagina & uterus with compression of adjoining structures consistent with hydrometrocolpos.

Cysto-vaginoscopy revealed a common urogenital tract. The hydrometrocolpos was drained and child later discharged home.

Fig 3





Re-Presentation

She re-presented at 6 weeks of age; unwell, vomiting, pale, tachycardic with poor weight gain. Her initial results demonstrated:

- **metabolic acidosis**
- **hyponatraemia (sodium 117mmol/L)**
- **hyperkalaemia (potassium 7.1mmol/L)**
- **abnormal renal function**
- **anaemia (haemoglobin 69g/dL)**
- **urine dip - microscopic haematuria only**

She was treated for a salt losing adrenal crisis with a 10ml/kg bolus of 0.9% sodium chloride, hydrocortisone, fludrocortisone. Hyperkalaemia was treated with IV bicarbonate, calcium gluconate and salbutamol. A blood transfusion was given as imaging showed a re-accumulation of the mass with internal haemorrhage. After stabilisation she had a cystovaginostomy, although only milky fluid was drained from the mass.

Further investigations demonstrated:

- **raised aldosterone and renin and cortisol**
- **17-hydroxyprogesterone and urinary electrolytes within the normal ranges.**

These results and resolution of signs following surgery demonstrate PHA.

Conclusion

PHA due to obstructive uropathy has been previously described, however, to our knowledge, there are no reports of obstructive uropathy secondary to hydrometrocolpos secondary to a common urogenital tract.

In PHA there is renal tubular dysfunction due to pressure from hydronephrosis and the release of intrarenal cytokines. There is an immature or resistant renal tubular responsiveness to aldosterone during infancy, leading to a picture with high circulating aldosterone levels but hyponatraemia & hyperkalaemia.

Clinicians should be aware of this uncommon presentation.

References
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2. Shee et al. A case of pseudohypoaldosteronism as the presentation of bilateral ectopic ureter in an infant. OMJ. 25, 137-140 (2010).

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