Prune-belly syndrome (Eagle-Barrett syndrome)
A Case Study

Judy Pullen RGN, ENB216
Clinical Nurse Specialist – Lead Nurse, Stoma Care
Great Western Hospitals NHS Foundation Trust
Swindon
Prune-Belly Syndrome (PBS)

- PBS is a rare genetic birth defect
- Aetiology unknown
- Affecting 1:40,000 births (Baird & MacDonald, 1981).
- Predominantly affects males (97%)
- Manifests as a congenital disorder of the urinary tract
Main clinical features

1. Failed abdominal wall development
Main clinical features

2. Cryptorchidism (undescended testes) in males.
Main clinical features

3. Multiple urinary tract abnormalities
In adolescent and adults the abdomen can become “pot bellied”
Case study

JP

• Born 1995 – youngest of 4 children
• Mum reports that she was offered termination at 30 weeks gestation.
  • AGE 2 - Left nephrectomy, transureterostomy and reimplantation of ureter.
  Small bowel bladder augmentation with Mitroffanoff.
  • AGE 5 – Commenced dialysis with insertion of right femoral fistula graft
• AGE 7 - Renal transplant
Case study – associated problems

- Speech and language delay
- Conductive hearing loss
- Increase weight for height
- Hypertension
- Hyperthyroidism—now resolved
- Hypermobility of joints
Case study

Mitroffanoff:
- Initially thick mucus – mum managed 3 hourly catheterizations during the day.
- Overnight free drainage
- Normasol bladder washouts

Gastrostomy:
- Overnight feeds

Schooling:
- JP had multiple absences
Referral to SCN

Referred in April 2015 – aged 19

Problems:

• Unable to catheterise Mitroffanoff since August 2014

• Urethral catheter

• A new Mitroffanoff would be a challenge!

Time to talk about urinary diversion with ileal conduit
Post operative

JP finally had surgery 4\textsuperscript{th} November 2015

• Inter operative problems resulted in a lengthy I/P stay

• JP underwent urinary diversion with ileal conduit.

• He developed adhesion obstruction – back to theatre but adhesionolysis NOT an option.
  “Bowel like tissue paper”

• Discharged home 5\textsuperscript{th} January 2016
Post discharge SCN review

• Attended stoma clinic with Mum in a wheelchair.

• Still had tunnelled central line should further TPN be required.

• Mum reported frequent UTI’s whilst JP informed me that he could “pee normally”.

• Had experienced mucus via urostomy and per urethra.
UROSTOMY

- Mum had to remind JP to change his pouch.
- Both preferred clear pouch.
- No leaks reported – urine was clear with minimal mucus.
- Skin clear
- No evidence of PSH, prolapse or retraction
Subsequent SCN review

- JP no longer using wheelchair but uses sticks for stability.

- Actively looking for 2 hours / day work and increase as tolerated.

- Oral fluid intake good.

- Taking Vitamin C – no mucus and urine clear.

- Still no PSH, prolapse or retraction of stoma.
Where are we now?

• JP continues to experience joint pains

Fertility:
• Unlikely to father children – only 1 undeveloped testicle

Prognosis:
• Generally poor in neonates and infants.
• 50% do not survive to age 2 (Duckett 1972)
• Early diagnosis and appropriate treatment prolongs life expectancy.
• A study of 50 patients revealed overall mortality was 32% (Burbige et.al 1987)
References


• Duckett JW. The prune belly syndrome. In: Clinical Pediatric Urology, Kelalis PP, King LR (Eds), Saunders, Philadelphia 1976

THANK YOU