The use of a two-piece ostomy pouching system in paediatrics, could it be a life changer?

Emily Counsell
Community Children's Nurse, New Zealand.

Introduction

The excitement and expectations of new parenthood can be quickly crushed when a newborn is diagnosed with a life-threatening condition which requires immediate surgery.

This case study will follow the journey of Rhys and his parents Jasmine and Michael after he was diagnosed with Mosaic Down's syndrome and Hirschsprung's disease.

In the hope that their experience may help others, Rhys's parents have consented to the sharing of their story. At their request, pseudonyms have not been used in this case study.

Mosaic Down's Syndrome and Hirschsprung's Disease

Down's syndrome is due to either extra genetic material or even a third copy of the chromosome 21 complex. It is the second most common human chromosomal disorder[1]. A wide variance in the characteristics of Down's syndrome presentation makes each child, born with this condition, totally unique[1].

Commonalities which can present included:

- The posterior of the head is flat in line with the neck
- A flat nasal bridge
- Epicanthal folds (a fold of skin in the inside corner of the eye)
- Almond shaped eyes
- A protruding tongue
- A single crease across one or both palms
- Short fingers with a single flexion on the fifth finger (two knuckles instead of three)
- Short toes and a large space between the first and second toes
- Short stature[2]

The life expectancy and social achievements of those born with Down's syndrome have risen significantly over the last two decades[3]. By 2002 the life expectancy of a child born with Down's syndrome in Australia was 60 years[3].

Mosaic Down's syndrome is diagnosed when there is a mixture of two different types of cell structures. Some cells will have the usual 46 chromosomes, and some will have 47. Children born with a mosaic mix of cells, some with 46 chromosomes and some with 47, can demonstrate fewer characteristics of Down's syndrome[4].

Approximately 5% of those born with Down's syndrome will have an associated digestive malformation[1].

The most common digestive malformations associated with Down's syndrome are imperforate anus 3% and Hirschsprung's disease 1%[1].

Hirschsprung's (HD) disease is characterised by the absence of parasympathetic ganglia in the distal bowel resulting in intestinal obstruction due to lack of motility[5]. More than a third of infants born with HD will require a stoma prior to definitive surgical resection[5].

Who is Rhys?

Rhys is Jasmine and Michael's 1st child. He was born at full term via a caesarean section. At two days of age, Rhys developed abdominal distension and bilious vomiting. An abdominal x- ray confirmed dilated loops of bowel indicating an obstruction. Following bowel biopsies, a diagnosis of Hirschsprung's disease was confirmed.

Within two weeks of birth, Rhys was also diagnosed with Mosaic Down's syndrome.

For Rhys's parents, who had no antenatal awareness of his conditions, the shock and anxiety must have felt overwhelming. Their expectations of new parenthood had been dramatically changed.

Rhys, lives with his parents and his 1-year-old brother Kyle. Jasmine is the family's principal income earner while Michael is primarily responsible for childcare.

PMHx

- At 10 days of age Rhys underwent a sigmoid colectomy with formation of an end colostomy
- At 9 months of age, he had a Soava procedure (see fig 1.) with reversal of colostomy. Following this procedure, he experienced abdominal pain and pain on defecation. He returned to surgery for formation of an end ileostomy with a mucous fistula.
- In 2020 a second attempt at reversal of ileostomy with parastomal hernia repair rapidly led to a return of rectal pain and re-formation of a new ileostomy, Rhys's third stoma.

Jasmine and Michael had sourced international review of Rhys future surgical options. There are provisional plans for surgery in the USA or Australia however due to COVID 19, these are on hold. If any future surgery does not achieve the desired outcome they will wait until Rhys is old enough to articulate his preference either for or against further surgical intervention.

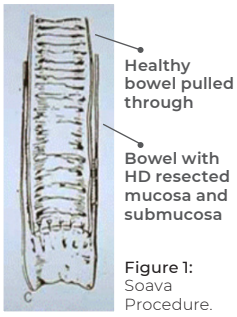

Figure 1: Soava Procedure.

Soava Procedure: Bowel where HD is present has the mucosa and submucosa resected. Healthy bowel is then pulled through into this section. This section of bowel forms a cuff around the healthy section which has been pulled through

Stoma Assessment

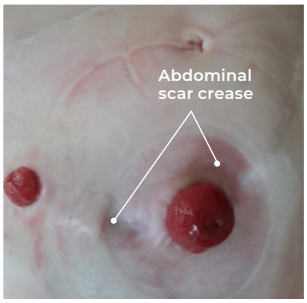

Figure 2: End ileostomy and mucous fistula show scarring and abdominal creases.

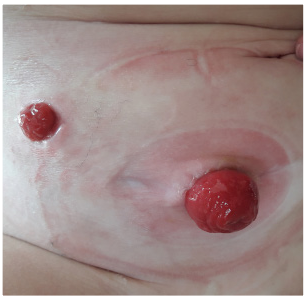

Figure 3: Rhys in the sitting position, skin slight red immediately after pouch removal.

Stoma Assessment

- **Skin:** Due to the incredible care from his parents and despite multiple leaks Rhys's skin was intact at this first review
- **Abdominal Contours:** Rhys has the expected child abdominal shape with a narrow, rounded contour
- **Parastomal contours:** In the sitting position, Rhys has a surgical scar crease at the 3 o'clock position directed towards his umbilicus, and another in the 9 o'clock position directed towards the mucous fistula
- **Mucocutaneous junction:** intact
- **Output:** normal consistency and volume

The Issue

Throughout Rhys's complex stoma journey there has been a plethora of issues in sourcing a pouching system which would meet his needs. While an acceptable pouch wear time in paediatrics is shorter than for adults, Rhys's parents have trialled five to six different pouching systems with various accessories however, none have achieved an acceptable wear time. At worse, they were changing the pouch up to six times per day. The stress caused by leaks, constant pouch changes, product reactions, the constant need to check the pouch for leaks, the variable deteriorating condition of Rhys's skin, and his increasing reluctance to participate in painful pouch changes have been very challenging for the family. The social isolation that resulted from the constant need to address Rhys's needs was testing on Jasmine and Michael's resilience.

Having trialled nearly all paediatric pouch ranges and with Rhys now outgrowing most of these, options were becoming limited. I reached out to Omnigon's Territory Manager and we agreed to do a community visit and assessment.

The Solution

Objective: To achieve an acceptable pouch wear time (at least 24 hrs.) which would allow this family social engagement and relief from constant pouch care.

Product Selection and Rationale

WELLAND ADHESIVE REMOVER WIPES

Rationale: The fragile nature of children's skin and their well-conditioned pain response to pouch changes has made remover wipes mandatory in my paediatric practice. The Welland Adhesive Remover wipes reduce trauma, pain (no alcohol), skin stripping and the damage which can be caused by pouch changes. Because the product does not need to be washed off and will not reduce the adhesion of a new pouch, it extended the opportunity to complete a pouch change before Rhys's abdomen became a moving target vanishing off at speed.

WELLAND HYPERSEAL® WASHER WITH MANUKA HONEY

Rationale: The leak point was the deep crease at the 3 and 9 o'clock which only became apparent when Rhys was in the sitting position. Although Rhys's skin at our first review was intact, he had a long history of eroded skin issues. The Welland HyperSeal® with Manuka honey was introduced to fill the crease and potentially prevent recurrence of the skin damage.

eakin dot® 2-Piece Baseplate and Pouch

Rationale: The eakin dot® 2-piece skinsmart™ technology has achieved predictable, secure, and extended wear times in adults, all of which we hoped to achieve for Rhys. This was my first use of the product on a child.

The eakin dot® hydrocolloid baseplate has a foam backing which is soft and flexible aiding in moulding to the abdominal contours. This was the feature that we needed to conform to Rhys's creased abdominal surface. It was also likely to increase his wearing comfort. The fluted edges of the baseplate allowed it to mould over the narrow-rounded body shape of a child.

We also needed a product that would stand up to everything that an active two-year-old could throw at it. We expected that Rhys would explore and challenge the coupling mechanism. The coupling mechanism on the eakin dot® proved resilient to withstanding these challenges.

Removal of the eakin dot® did not leave hydrocolloid residue on Rhys skin which facilitated faster pouch clean ups when changing.

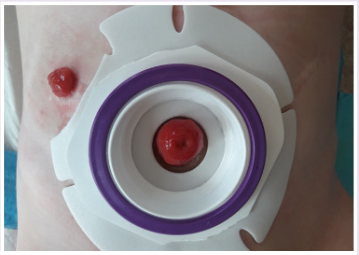

Figure 4: Rhys with eakin dot® adapted to accommodate the mucous fistula and not rub on his thigh when sitting. Fluted baseplate edges have moulded over rounded belly.

The oval shape of the eakin dot® baseplate allows the baseplate to be turned having the longer axis either vertically or horizontally depending on the shape of the abdomen. This gave us more options to custom fit the baseplate to Rhys's abdominal shape.

The Outcome

Our initial goal of achieving a 24-hour wear time for Rhys has been exceeded with the eakin dot® 2-piece. He now has an achievable wear time of three days. The impact of this has been far reaching.

Rhys is now three years old and has been using the eakin dot® 2-piece for a year. Rhys attends kindy with his younger brother four days a week. While slightly delayed in achieving some milestones, Rhys is doing exceptionally well at kindy. He is sociable, keen to participate in all activities and to explore his world. Attending kindy had not been possible prior to the use of eakin dot®. Overall, the use of eakin dot® has allowed this family to lead a more normal life and to socialise outside their home.

Conclusion

From my experience in caring for Rhys and his family, I take forward validation of my existing practice, new knowledge, and a deeper awareness of the psychological impact that repeated appliance failures can have on a patient and their family. In sharing the impact that these failures had on their ability to cope, socially, psychologically, and emotionally, Jasmine and Michael have made me more aware of how critical the right pouch at the right time is. For them this was the eakin dot® 2-piece.

Since using the eakin dot® with Rhys, I have successfully replicated its use with other paediatric patients. I now have the confidence to explore beyond the obvious choices in product selection and employ the wider network of ostomy specialists to help create better patient outcomes.

It has been a privilege to be involved in Rhys's care. I remain in awe of his parents, Jasmine and Michael, and thank them for allowing me to share their experience so health professionals can learn and improve practice.

The final words belong to Rhys parents "Love the bag the only bag (we) truly had success with".

References

1. Bermudez Americam Journal Medical genetics 2019, Aug 17: 9(8) 1426-31
2. Stanford Children's Health – Lucile Packard Children's Hospital Stanford " Mosiac Down Syndrome" -2021 Stanford Children's Health - [cited 2021 April 20] . Available from: https://www.stanfordchildrens.org/en/topic/default?id=mosaic-down-syndrome-90-P02133
3. Holmes,G. Gastrointestinal Disorders in Down Syndrome: Gastroenterology and Hepatology from Bed to Bench. 2014:7(1):6-8
4. International Mosaic Down Syndrome Association. [Cited 2021 April 14] Available from: https://www.imdsa.org/characteristics/