# Epidermolysis bullosa: a rare challenge for peritoneal dialysis nurses

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## Abstract

Epidermolysis bullosa is a genetic skin disorder characterised by blister formation in response to minimal trauma or friction. Our renal service was required to provide peritoneal dialysis (PD) for a young lady suffering from this rare disease and this paper provides an insight into the way in which we collaborated with the health care team and the patient and family to provide optimum dialysis management to suit the patient's specific clinical needs.

The key challenges we faced included the placement of a suitable PD catheter and the subsequent exit site care. The patient had multiple skin lesions and required daily baths and special dressings to most of her body. In addition, we aimed to provide optimal dialysis with reduced fill volumes as although she was 24 years old she had the body frame of a young child.

Communication and collaboration with the patient, family and a variety of medical teams involved in her care were critical to the success of PD. Individualised care and tailored PD therapy is necessary for every patient, but more so as we encounter patients with special conditions/needs.

## Keywords

Peritoneal dialysis, epidermolysis bullosa.

## Introduction

The aim of this paper is to provide awareness and insight into the rare skin condition epidermolysis bullosa (EB) and the challenges faced by our peritoneal dialysis (PD) service in managing a patient with EB and advanced chronic kidney disease (CKD).

EB is an umbrella term for a group of rare genetic skin disorders characterised by blister formation in response to minimal trauma or friction (Pillay, 2008). The onset of EB is at birth or shortly after, with at least 30 different phenotypes of inherited EB described in the literature. These phenotypes occur as a result of molecular mutations within gene encoding for structural proteins within the epidermis or skin basement membrane zone (Fine & Mellerio, 2009). There are four major types of EB: epidermolysis bullosa simplex; junctional epidermolysis bullosa, dystrophic epidermolysis bullosa and a mixed type called Kinder syndrome. This typing of EB is based on ultrastructural differences noted by transmission electron microscopy (Fine & Mellerio, 2009).

The severity of EB ranges from mild to lethal, depending on the EB classification. Many of the more severe subtypes of EB are associated with extracutaneous complications including chronic anaemia, osteoporosis and gastrointestinal and cardiac involvement. Epithelial surfaced tissues at particular risk include the external eye, upper airway, oesophagus and genitourinary tract. Table 1 presents an overview of the various manifestations of EB including both skin and extracutaneous sequelae. Some of these manifestations may be life-threatening even in early infancy (Fine & Mellerio, 2009).

In Australia there are approximately 1,000 mild to severe cases of EB and in New South Wales there were 140 people enrolled on the EB Registry in 2010 (Murrell, 2010).

Currently there is no definitive treatment for EB. Clinical management is mainly supportive, focused on preventing and treating blisters and infections through close monitoring, daily baths and specific dressings. Eye and oral care are essential, as lesions may appear in these areas as well. Pain during bathing and dressing change can be substantial and pain relief is essential prior to these activities. In all cases, patients with EB must consume high-caloric and protein-fortified foods and beverages to help replace protein lost in the fluid from draining blisters. In severe cases, oesophageal lesions may be present, leading to poor appetite, chronic nausea and vomiting; therefore, enteral or parenteral supplementations may be required (Denyer, 2010; Abercrombie, Mather, Hon, Graham-King & Pillay, 2008).

EB subtype specific differences exist in both the time of onset and severity of many of the extracutaneous complications.

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*Table 1. Epidermolysis bullosa – clinical presentations, complications and management*

| | Symptoms | Complications | Management |
|---|---|---|---|
| Ophthalmic<br>Fine & Mellerio (2009a) | Corneal abrasions, dryness and ulcerations | Pain and photophobia | Analgesia, darkened room and lubrication |
| Cardiac<br>Fine & Mellerio (2009b) | Cardiac myopathy | Increased cardiac death risk | Monitor nutrition, regular echocardiogram, urgent treatment of anaemia or iron overload |
| Gastrointestinal tract<br>Fine *et al.* (2008) | Blistered anal margins, dental problems and dysphagia | Constipation, gastric reflux, malnutrition and microstomia | Dental treatments/implants, enteral supplementation, laxatives, oesophageal dilatation and proton-pump inhibitors |
| Genitourinary tract<br><br>Chan *et al.* (2006) | Eroded groin or vulval areas, glomerulonephritis, renal amyloidosis, immunoglobulin A nephropathy and obstructive uropathy | Chronic renal failure<br><br>Sexual inactivity | Renal replacement therapy<br><br>Hormone therapy |
| Skin<br>Fine & Mellerio (2009a) | Extensive blistering and chronic wounds | Lack of intact skin and skin infections | Drain blisters, bland emollients, oral anti-pruritus, special dressings (non-adherent and soft silicone) and nutritional supplementation |
| Muscular<br>Pillay (2008) | Contractures due to atrophic scarring | Pseudosyndactyly or "mitten deformity" | Surgery |
| Haemopoietic<br>Pillay (2008) | Blood loss from wounds and poor iron intake | Chronic anaemia | Iron supplements |
| Skeletal<br>Martinez & Mellerio (2010) | Increased bone turnover | Osteoporosis and osteopenia | Annual energy x-ray absorptiometry scans, exercise, calcium and vitamin D supplements |
| Sensory<br><br>Pillay (2008) | Multi-factorial pain: wounds, contractures, bones, dental, GIT, procedural and corneal ulceration | Chronic and generalised pain | Ascertain cause of pain and pain team referral |
| Malignancy risk<br>Fine & Mellerio (2009)a | Squamous cell carcinoma | Short life span | Surgery, grafting, radiotherapy and chemotherapy |
| Psycho-social<br><br>Pillay (2008) | Anxiety, depression and non-compliance (patient)<br>anger, confusion grief, guilt and financial hardship (family) | Emotional, financial and social distress (patient and family) | Counselling and education.<br>Psychological and pharmacological treatment.<br>Community and philanthropic support. |

Detailed understanding of the breadth and clinical impact of these complications, and which EB subtypes are at highest risk for their development, will facilitate surveillance and early medical or surgical intervention.

As highlighted, there is no definitive cure for EB but preliminary clinical data in both mouse and human models have demonstrated the feasibility of bone marrow transplantation, showing longer term survival and reversion of the phenotype with normalisation of the skin basement membrane and the formation of anchoring fibrils (Kiuru *et al.*, 2010).

## Patient background – health history and assessment

In August 2011 our PD staff were asked to perform an urgent pre-PD assessment and provide education for a young female patient admitted to the renal ward. To maintain this patient's anonymity we have called her Sam.

The verbal referral briefly stated that Sam had EB and was diagnosed with CKD in 2010. There had been a rapid decline in renal function and she required the urgent surgical placement of a PD catheter and immediate commencement of dialysis. Although her blood results indicated the need for urgent dialysis, haemodialysis was not considered due to poor venous access and poor skin integrity.

Sam suffers from the more severe form of EB, which is recessive dystrophic; characterised by blisters over large body surfaces, loss of nails, fused fingers and toes, atrophic scarring, itching, anaemia, growth retardation, plus blistering and scarring, inside the mouth and gastrointestinal tract, causing chronic nausea and malnutrition (Fine & Mellerio, 2009). Significant renal and urological complications have been reported in patients with recessive dystrophic EB. Renal failure has been reported in two paediatric patients (Chan, Dillon, Duffy & Atherton, 2007) and in a 29-year-old female (Ahmadi & Antaya, 2007). These cases were treated with PD.

Sam is a 24-year-old graphic designer who lives with her parents. Her parents assist or attend to most of her care and daily living activities due to her frailty, fused fingers and poor mobility. On interview she was alert and orientated. At assessment she was able to walk only short distances and had a child-like voice and body-frame. Her body and her extremities were well covered, with bandages and dressings, due to generalised skin lesions and blistering from EB. On closer examination, her abdominal and chest region lacked healthy and intact skin. In addition to EB, her medical conditions included osteoporosis, hyperparathyroidism, fatty liver and advanced CKD.

The following are the main challenges we faced in providing PD in a patient with EB.

## Insertion of the catheter

Sam required urgent transfer from the ward to ICU for stabilisation due to uraemic seizures, coffee-ground vomit and malaena. She was tachycardic and her Hb dropped to 52 g/L. She was intubated and sedated in ICU until she was clinically stable to undergo PD catheter insertion. Surgery was rescheduled for the following day and plans made to determine the type of PD catheter that could be used. Due to Sam's small size she required a paediatric-sized catheter. There was a lack of intact skin surrounding her abdominal area and she required daily baths as a critical component of her routine skin care.

A PD catheter was placed pre-sternally where the surgeon was able to locate the healthiest skin and an adult-sized catheter was cut to size instead of using a paediatric catheter. After surgery, Sam continued to be intubated and sedated for 11 days, which gave the PD staff the opportunity to establish PD without causing discomfort or pain.

## Catheter care

The next challenge following the PD catheter insertion was her postoperative PD catheter exit site care. The department's exit site dressing policy was not appropriate due to the need to use special dressings and tape on Sam's sensitive, blistered skin. In general, Sam's body dressings were changed daily by her parents and reviewed by the dermatology team regularly. Our aim was to reduce the risk of exit site infection while maintaining skin integrity, notwithstanding the fact that Sam had chronic skin wounds and she was colonised with a number of organisms including Klebsiella in her blood and multi-resistant *Staphylococcus aureus* on her lower and upper extremities, skin folds and torso.

People affected by severe forms of dystrophic EB may require extensive dressings in an attempt to heal wounds and to offer protection against friction and shearing forces (Denyer, 2010). Considerable pain is felt by the patient during dressing changes and pain relief is often required (Pillay, 2008). We worked closely with the dermatology team and Sam's parents to establish her postoperative exit site care and dressing regimen. Postoperative dressing care consisted of daily Betadine swabs and Excilon™ AMD™ (Tyko Healthcare, USA) antimicrobial sponge dressings secured with Mepitac tape® (Mölnlycke Health Care, Sweden). Sam's parents were vigilant in not letting staff remove or replace the dressings without notifying the PD nurses. ICU nurses were educated on the importance of keeping the PD catheter immobile and limiting access to the PD catheter exit site. We educated selected renal ward nurses on the daily postoperative dressing regimen for after-hours support.

The collaboration between the patient, her parents, nurses and medical teams was a hallmark of Sam's care and proved to be very effective as Sam did not experience an exit site infection throughout her prolonged stay in ICU, despite pre-existing skin infections. Of all the skin swabs taken, only the PD catheter exit site swabs showed no growth.

The next step in PD catheter management was to establish a suitable long-term exit site dressing regimen. Considerations included her need for daily baths, complex body dressing regimens and her vulnerability to recurrent skin infections due to recurrent skin blistering and lesions. Ahmadi and Antaya (2007) described their experience with providing PD for a patient with EB. PD catheter exit site dressings consisted of a simple, non-adherent dressing and the patient's underpants were used to maintain the dressing in place. The reported outcome at one year was that there was no blistering or any skin disease around the catheter. Given the success of our postoperative exit site care and dressing, we were inspired to continue the daily Excilon™ dressings; however, they were quite expensive. With the help of Sam's parents, we devised a new dressing regimen. A product called Mepilex Border® (Mölnlycke Health Care, Sweden) has similar bacteriostatic qualities of an Excilon and does not require daily replacement because it is essentially waterproof. The product is supplied by the same company as Sam's body dressings and the Dystrophic Epidermolysis Bullosa Research Association Australia (DebRA) was happy to include Mepilex Border®, as part of Sam's regular dressing supply. Sam's parents were instructed to change the PD catheter dressing daily, then second daily. Currently she has third daily dressing changes as her parents discovered the dressings remained waterproof for at least three days.

## Providing safe and effective dialysis

Due to Sam's small body size we had to determine an appropriate PD fluid fill volume. The advice from the paediatric nephrologist was to start with 10 ml/kg (approximately 250–300 ml fill volume). We were unsure as to whether this volume would provide Sam with enough dialysis to prevent further uraemic seizures. Long-term fill volumes were also a challenge as we aimed to optimise her dialysis regimen without causing too much intraperitoneal pressure which could precipitate to more nausea and vomiting episodes.

Automated intermittent peritoneal dialysis (AIPD) was commenced immediately postoperatively, which proved problematic, with frequent alarms due to negative ultrafiltration and PD fluid retention. As we increased Sam's fill volume the flow rate improved causing fewer alarms although when AIPD was running and Sam was alert, she complained of abdominal pain mostly during the drain cycle. Sam's PD prescription was then changed to an 85% tidal program and we gave her oral analgesia. Her nephrologist trialled her on intraperitoneal lignocaine. We administered 1% lignocaine (50 mg) in each 6 Litre dialysis bag before dialysate infusion. These combined measures helped alleviate her abdominal pain on dialysis.

As was anticipated, the low-fill AIPD was suboptimal for Sam's initial dialysis needs and a paediatric femoral vascath was inserted. Continuous venous to venous haemodialysis was performed while we persisted with PD until the appropriate fill volume of 500 ml was established. Sam's dialysis prescription on discharge from the hospital was an eight-hour APD therapy of 500 ml fill volumes with no day dwell. Over the next six months this fill volume progressively increased up to 900 ml and then a day dwell of 600 ml was introduced.

## Additional challenges

During her stay in hospital, Sam's body weight dipped to only 27 kg and she consistently complained of nausea and vomiting. Dysphagia, oesophageal strictures and a number of other gastrointestinal manifestations are well-known complications of dystrophic EB (Fine, Johnson, Weiner & Suchindran, 2008) and for Sam gastroscopy revealed that the EB had affected her oesophagus. Sam was trialled on a small dose of dexamethasone, to which she responded well and she received both oral and enteral feeds during her hospitalisation to ensure she was receiving adequate nutrition.

During the completion of this paper, Sam presented to clinic with pain along the PD catheter tract. Ultrasound revealed a collection along the PD catheter tunnel. Unfortunately after nine months the catheter was removed and she has since commenced haemodialysis via a tunnelled cuffed catheter. The PD catheter tip was cultured and was positive for *Klebsiella*. The provision of haemodialysis treatments for Sam is presenting its own challenges but we are cognisant of the disease process and special requirements of Sam regarding dressings and this has helped the haemodialysis staff. At this stage, we are planning to insert another PD catheter after six weeks on haemodialysis

## Conclusion

On discharge, Sam's nausea was well controlled with Dexamethasone and her appetite improved. She weighed 31–32 kg with an albumin level of 17 g/L, a marked improvement from 27 kg body weight and albumin of 10 g/L. Sam's serum biochemical markers were stable with a Hb maintained between 70 and 80 g/L. Her most recent Kt/V (1.51) and creatinine clearance (44.19 L/week/1.73 $m^2$), were less than our targets Kt/V >1.7 and creatinine clearance >60; therefore, we increased her to an APD fill volume of 900 ml with a day dwell. To date she has not experienced a PD catheter exit site infection or peritonitis episode and, most important of all, is that she feels well and is back to her usual activities including graphic design.

We have aimed to present the challenges of providing dialysis treatment for a young lady suffering from the rare disease EB. Through determination and ingenuity we were able to resolve Sam's PD issues one by one, successfully providing PD. Teamwork patience and good communication between the renal team, dermatologist and Sam and her family were critical to the success of PD.

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