Epidermolysis Bullosa: A Retrospective Analysis in Radiation Dose and Fluoroscopic Techniques.

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Learning objectives

The learning objectives for this retrospective analysis were:

1. To identify the cohort of patients with Dystrophic Epidermolysis Bullosa (DEB) at Guy's and St Thomas' Hospital who have undergone fluoroscopic procedures.

Data was collected retrospectively within the fluoroscopy department using the Computerised Radiology Information System (CRIS) and Picture Archive Communication System (PACS) over a two year period, from April 2012 to April 2014.

2. To record the following parameters in each of the patients:

   • Patient age
   • Patient gender
   • Classification of DEB (recessive or dominant)
   • Total number of swallows and oesophageal dilatations
   • Number of formal contrast swallow examinations
   • Total screening time, DAP and skin dose
   • Oral contrast used
   • Findings/stricture location
   • Oesophageal levels involved (single or multiple)
   • Number of acquired views during each study
   • Did the stricture location alter (if the study was repeated)?

The main focus was on patient radiation dose, the pathological findings and the method of study acquisition.

3. There is a tendency for pharyngo-oesophageal strictures to recur in the same place. (7) By altering our fluoroscopic methods, such as carrying out targeted studies and reducing the number of acquired views, this may reduce exposed radiation dose.
Background

Epidermolysis Bullosa (EB) is an inherited connective tissue disorder with a UK incidence of one in 17,000 live births. It is characterised by blistering of the skin and mucous membranes in response to mechanical trauma (1). Patients who present with dysphagia and malnutrition secondary to gastro-intestinal mucosal involvement and strictures tend to be those with Dystrophic Epidermolysis Bullosa (DEB).

DEB constitutes a group of diseases caused by defects of anchoring fibrils, caused by mutations within the gene encoding type VII collagen, COL7A1. Type VII collagen is a large macromolecule which dimerises to form a semicircular looping structure or anchoring fibril. As a consequence of these mutations, blisters heal followed by dystrophic scarring.

DEB is divided into two main groups characterised by the inheritance pattern - dominant and recessive. In recessive DEB (rDEB) forms, COL7A1 mutations usually cause premature termination codons, resulting in an absence of type VII collagen in tissue, which tends to signify more severe disease (2).

Involvement of the internal mucosa can result in microstomia, oesophageal strictures and webs, urethral and anal stenosis, corneal scarring and phimosis. Affected individuals are often severely malnourished due to trauma to the oral and oesophageal mucosa, and require multiple oesophageal dilatations and feeding tubes for nutrition (1-3).

Guys and St Thomas’ Hospital in London is one of two national centres in the UK for the diagnosis and clinical care of patients with EB (5). The focus of this retrospective analysis was on the formal fluoroscopic examinations carried out in our diagnostic department prior to intervention. As DEB patients tend to be younger and prone to having multiple studies, it has been suggested that we be more vigilant of their radiation exposure and carry out targeted studies (6, 7).
Findings and procedure details

Between April 2012 and April 2014, 22 patients underwent a total of 93 contrast swallows and 47 oesophageal balloon dilatations, with an average of 4.2 swallows and 2.1 dilatations per patient, the equivalent of two swallows and one dilatation per year per patient. The total number of pre-procedural contrast swallows carried out in the fluoroscopic department at St Thomas' Hospital was 55, with the number of check swallows (post-procedure) being 38, which fell short of the 47 dilatations undertaken.

Patient Demographics

The mean age of the patient was 38 years, with the youngest being 20 and eldest 71 years of age. There was a female to male preponderance of 68% to 32%. Classification of DEB showed 66% had the recessive (rDEB) and 34% had the dominant phenotype (dDEB). The most popular contrast used was Barium Sulphate (Baritop) solution rather than a water soluble contrast.

Radiation Parameters

The total screening time was calculated as the sum of all the individual formal contrast swallow studies undertaken within the fluoroscopic suite at St Thomas' hospital. The maximum total screening time was 5 minutes and 4 seconds in a patient who had undergone the most number of formal swallows in a two year period (eight in total) and the minimum screening time was 10 seconds. The mean screening time was 1 minute 19 seconds per patient.

The Dose Area Product (DAP) ranged from a total of 69.3 to 3697.89 Gycm² per patient. The mean total DAP equalled 775.7 Gycm². There was a direct correlation with the screening time and skin dose. Peak skin dose (measured in Gy) is a measure of the likelihood of radiation induced skin effects, which is of particular concern in DEB patients. It ranged from 5.87 to 178.57, with an average dose of 41.7 Gy. (Fig.2)

Pathological Findings

To classify each of the strictures into an oesophageal division, the American Journal of Cancer Committee's (AJCC) divisions was used (8):

• Superior to oesophagus/oropharynx (Above C6)

• Cervical oesophagus (C6-T3): cricopharyngeus to thoracic inlet
• Upper thoracic oesophagus (T3-T4): thoracic inlet to carina
• Middle thoracic oesophagus (T5-8): carina to inferior pulmonary vein
• Lower thoracic oesophagus and GOJ (T9-10)

In total, 34 strictures were identified, many of which recurred. With regards to pathology seen, 84% of patients had oesophageal or pharyngeal strictures, with 14% of patients having a lone or associated pharyngo-oesophageal web. (Fig.3) One patient was shown to have a lone pharyngeal pouch, whilst two patients had hiatus hernias. Fifteen of twenty-two patients (68%) underwent oesophageal dilatation. In patients with strictures, 40% had involvement of one oesophageal level, whereas 60% had involvement of two or more levels.

If the stricture(s) recurred, was there any difference in anatomical location, when compared to previous studies? Out of 22 patients, only sixteen were eligible in assisting us in answering the above question. In 94% of patients the position of the stricture(s) did not alter. Only in one patient did formal swallows show a difference in the position of the stricture (previously the cervical oesophagus to the upper-middle thoracic oesophagus).

Acquisition by Radiologists

Out of a total number of 55 formal swallows, the number of views of the oropharynx and oesophagus acquired by the radiology speciality registrar conducting each fluoroscopic examination were measured, with almost half (47%) of all examinations adopting a four-view technique (as shown below). In 27% of studies, three views were acquired, 18% used a two-view and 7% a one-view method of acquisition.

• Oropharynx/cervical oesophagus: lateral
• Oropharynx/cervical oesopahagus: antero-posterior
• Oesophagus and gastro-oesophageal junction: antero-posterior
• Oesophagus and gastro-oesophageal junction: right anterior oblique

Targeted studies, (two views or less) were carried out in only 25% of formal fluoroscopic studies. They focused on previously noted areas of stricture formation. This was shown to expose the patient to less radiation.
Fig. 2: Radiological parameters recorded for all patients including total screening time, DAP and skin dose.

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**Fig. 3:** Pathological fluoroscopic findings and their anatomical location.

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Fig. 4: A four view fluoroscopic study on a rDEB patient who had undergone 8 formal fluoroscopic contrast swallow tests in 2 years. Note is made of the oesophageal stricture at the T3-T4 level and the double RAO view of the oesophagus.

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Fig. 5: The same rDEB patient has a targeted study, with a single RAO view. The stricture has re-presented in the same location.

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Conclusion

In this retrospective analysis, the most common fluoroscopic pathological findings were stricture within the oropharynx and cervical oesophagus. Sixty percent of patients with a stricture, had multiple strictures in 2 or more divisions. The majority of strictures did recur in the same position (94%).

Our methods of fluoroscopic acquisition tend to show that we undertake thorough fluoroscopic examinations on often young DEB patients who have a similar recurrence of their pathology.

Radiation dose in DEB patients was related to the number of studies carried out and the number of views acquired. Those with a recessive phenotype were more likely to have severe disease and therefore more examinations.

If the anatomical location does not alter, should we be undertaking targeted studies in all our patients, rather than in only 25%? Would we be at risk of missing pathology (ie. a new stricture or malignancy) if we did?

It appears as though targetted views of a known recurrent oesophageal stricture would be appropriate rather than routinely obtaining standard antero-posterior and lateral views of the pharynx and right anterior oblique (RAO) views of the oesophagus in each case.

It is also important to consider that this is an analysis of the data to hand, with a focus on changing our practice. We require a larger cohort, over a longer period of time in order to assess the true significance of these findings.
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