(SIE); or as a further, poorly characterised form of AD skin peeling, termed epidermolysis bullosa simplex superficialis (EBSS), previously described in two families.

We report 6 affected individuals from 2 generations with generalised AD skin peeling. All presented neonatally with erosions at trauma-prone sites including the axillae, back and thighs, with ongoing skin fragility caused by friction. All 4 affected children reported slightly dry skin in the first decade of life with very mild hyperkeratosis of the axillae and neck. Some individuals had peeling of fingertips and soles, and one adult had mild diffuse planter hyperkeratosis. There was no erythema, mucosal, nail or hair involvement. Initially, EBSS was considered based on AD inheritance, the generalised distribution, and lack of inflammation and ichthyosis at presentation.

A biopsy of rubbed, uninvolved skin from one affected individual showed a thickened stratum corneum but no signs of blistering or intrastructural abnormalities at the dermal-epidermal junction or within the epidermis. Sequencing of KRT5 and KRT14 (keratin 5 and 14) showed no mutations, but whole exome sequencing demonstrated a heterozygous missense mutation in KRT1 encoding keratin 1, p.Ser338Pro, in the 4 probands tested. This amino acid substitution is located within the L12 linker region, close to where other pathogenic mutations in keratin 1 have been reported in unrelated individuals with EI. Therefore, the most likely diagnosis in this family is EI due to a novel mutation in KRT1.

This clinically mild disorder and new KRT1 gene pathology extends genotype-phenotype correlation in EI and underscores the value of next generation sequencing in diagnosing clinically atypical genodermatoses.

### References


### Abstracts

#### G29 A CASE OF PHYLLOID HYPOMELANOSIS – A RARE BUT SPECIFIC PRESENTATION OF CHROMOSOMAL MOSAICISM

**Aims**

Paediatric postgraduate training needs to prepare paediatricians for the future delivery of high quality care. Doctors in Postgraduate Training (DrPGT) are often best placed to identify safety/quality concerns and can innovate across organisational boundaries. To address this, a programme was developed at a large tertiary centre providing a supportive educational environment. Its aims are to allow experimental learning on an improvement project alongside teaching of quality improvement (QI) and systems theory.

**Methods**

EQuIP (Enabling Doctors in Quality Improvement and Patient Safety) supports DrPGTs through a QI project within their department, aligned to Trust’s objectives. A three level approach to the programme ensures DrPGT engagement. All DrPGTs participate in a 1hr workshop to understand the importance of QI (level 1). Level 2 is a 6-month rotational programme with 2 full day workshops on improvement methodology, project surgeries facilitated by managers, and mentoring with senior clinicians. Level 3 is more intensive, over a 9 month period, to develop expertise and deliver level 1 workshops. The innovation involves a peer-designed programme while being work-based, delivering organisational strategies. Pre- and post- programme questionnaires allow Kirkpatrick 4-level evaluation.

**Results**

All 40 participants agreed that the project was a valuable learning experience and that the programme met their expectations (level 1, reaction). Level 2, learning, was demonstrated by an improvement in QI definitions post programme, awareness of QI resources and confidence in using methodologies including PDSA and process mapping (P < 0.001). Post programme, all but one participant said they are planning another QI project and that they are more aware of improvement work in their unit (behaviour change, level 3). Benefits to the organisation (level 4) are evident from successful projects presented to the executive team showing reduction in DNA rates, improved theatre efficiency, improved quality of medical notes etc.

**Conclusion**

EQuIP changes the way DrPGTs view healthcare as they become quality champions for their department. The design and evaluation of EQuIP may inform similar educational programmes in other organisations. This capacity building is crucial to ensure future Paediatric leaders have the skills and motivation to improve the effectiveness of our healthcare system.
and to try translated versions of the tool to maximise the participation of respondents with limited knowledge of English in a diverse ethnic population.

**Background** Patient/parent satisfaction surveys are important monitoring tools used in the national health service (NHS). Before the Urgent-and-Emergency-Care PREM tool was published by the RCPCH in October 2012 there was no standard feedback form for paediatric A&E. This new tool is a well researched and standardised monitoring tool used in the national health service (NHS). Before the publication of the Urgent-and-Emergency-Care PREM tool was published by the RCPCH in October 2012 there was no standard feedback form for paediatric A&E. This new tool is a well researched and standardised monitoring tool used in the national health service (NHS).

**Methods** Demographic data from the census was collected which showed that our NHS trust caters for a population with a large percentage of Turkish/Greek Cypriot, African and Somaliani ethnicity. There is an annual attendance of about 38000 to children’s A&E. While doing the survey, we eliminated the bias due to language barrier by translating the RCPCH tool in Turkish and Somali, the two commonly spoken languages in our ethnic population-group. The PREM tool was translated by doctors with knowledge of the languages and was colour coded for adult or children versions. Questionnaires were given to consecutive willing parents and/or children while waiting in the department. The feedback forms were analysed on Microsoft Excel using common statistical methods.

**Results** Total of 50 feedback forms were collected. Feedback was given by 12(24%) children, 29(64%) parents and 4(9%) by both. The study group had 19(41%) European, 9(20%) Asian, 12(26%) African and 2(4%) mixed. The main languages spoken were English 25(57%), other European 10(22%), and all other 10(22%). The main highlights of the survey was that 43 of 46 (94%) respondents were satisfied by the services, 24(49%) waited longer than expected, 15(32%) wanted better information while waiting and 4(8%) were not given adequate privacy.

**Conclusions** The introduction of feedback forms in multiple languages has perhaps given a more unbiased feedback with more involvement of the ethnic subgroups. The results were overall satisfying but a few specific areas that need improvement were identified. The responses to individual questions will also serve as a baseline for serial monitoring after implementing changes and training in problem areas.

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**Abstract G32**

**DEVELOPING PRINCIPLES FOR DELIVERING AND COMMISSIONING BETTER HEALTH OUTCOMES AND EXPERIENCES FOR CHILDREN AND YOUNG PEOPLE SO THEY ARE COMPARABLE WITH THE BEST IN THE WORLD**

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**G32**

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**Abstract G32 Figure 1**

**Stillbirths Figures Mbarara Hospital 2012**