

# **Specialist and transitional care provision for Amelogenesis Imperfecta: a UK wide survey**

## **Authors and contributions**

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## **Abstract**

### Background

Amelogenesis Imperfecta (AI) can be challenging to manage, due to the complexity and variation of presentation. Clear care pathways between general practice, specialist Paediatric Dentistry and adult services are required.

### Aim

To assess the provision of specialist care and transitional care arrangements for paediatric patients with AI in the UK.

### Method

An online survey was disseminated to members of the British Society of Paediatric Dentistry in January 2020. Descriptive analysis was used to interpret the quantitative and qualitative results.

### Results

- 115 clinicians across all four nations participated.
- Most respondents (54% n=66), were based in the Hospital Dental Service.
- Overall, 29% (n=33) were Consultants and 24% (n=28) Specialists in Paediatric Dentistry.
- The most common patient age group seen was 6-12 years-old.
- No clear AI referral pathway into specialist care was reported by 49% (n=47).
- A clear transitional care pathway was deemed not to exist by 77% (n=72), with 85.9% (n=73) indicating a need.
- Qualitative analysis themes included: 'Unclear care pathways,' and 'Specialist care access problems.'

### Conclusion

Access to specialist paediatric dental care and transition to adult services is not readily available throughout the UK for AI patients. There is a clear need to establish and improve existing pathways.

## **Research in Brief**

- Access to specialist paediatric dental care and transition to adult services is not readily available nationally for AI paediatric patients.
- There is a need for to establish and improve care pathways for paediatric AI patients throughout the UK.
- There is enthusiasm in the UK Paediatric Dentistry workforce for improvement to AI care pathways to be made. Education, addressing workforce shortages and refining pathways are areas that require further development

## Introduction

Paediatric patients with Amelogenesis Imperfecta (AI) often require a combination of specialist dental input and care provision in general dental practice. The role of the General Dental Practitioner (GDP) is to provide routine care and identify those patients who require further assessment by a specialist team. This amalgamation of care should be available locally to families and include a smooth transition to adult services.

AI is a rare, genetic condition affecting dental enamel with a prevalence of 1 in 700 to 1 in 14,000 depending on the population studied.<sup>1</sup> The impact of AI is significant, and includes functional, aesthetic and psycho-social problems, in addition to difficulties providing effective treatment.<sup>2,3,4</sup> There has recently been documented a high burden of care for paediatric patients with AI in the UK NHS with improvement in care for this patient cohort essential.<sup>5</sup> In addition, a newly developed Patient Reported Outcome Measure (PROM) for this condition demonstrated from the patient's perspective, the range of problems, concerns and impact AI has on a child.<sup>6</sup> Opportunity to access early specialist assessments will improve management and aid transition of these patients to adulthood with comfortable, functional dentitions.<sup>7</sup> Robust relationships and clear pathways between specialist centres and primary care can ensure children and young people (CYP) receive comprehensive, timely care. The pathway should also be dynamic and allow variations such as patients transitioning from primary care to adult specialist centres whether or not Paediatric Dentistry had been involved.

Access to specialist paediatric dental care in the UK is varied. There are workforce shortages and geographical challenges with some areas lacking access to specialist paediatric dental teams.<sup>8,9,10</sup> This results in some areas of the UK where primary care clinicians may have difficulty in obtaining local specialist care access for patients. The challenges in providing treatment for these patients does not cease on reaching adulthood. There is a need for the paediatric team to have clear pathways for transition of these patients to adult services. Patients who are discharged from paediatric services into primary care alone may face challenges in referral acceptance back into specialist care as an adult. There may then be a financial burden put on these patients seeking care in the primary care setting and the GDP left unsupported in managing these patients resulting in an overall increased burden of care. Inequalities and barriers to transitioning paediatric patients to specialist adult dental services are required to be defined and explored before improvements can be made.

The aim of this study was therefore to assess the provision of specialist care and transitional care arrangements for paediatric patients with AI in the UK.

## **Materials and methods**

An online survey was disseminated by email to members of the British Society of Paediatric Dentistry (BSPD) in January 2020. A two-month period was allowed for completion. Further reminder emails were sent to both the consultants in Paediatric Dentistry and trainee groups.

The survey was designed by members of the AI Clinical Excellence Network (CEN). This included three consultants in Paediatric Dentistry from different units in the UK and with backgrounds from both the hospital and community service. The AI CEN was established in 2019 with an aim to promote clinical excellence for CYP with this condition. As part of this remit, this survey was devised to identify current care practices and aid with future project design and work-streams for the group. The questionnaire did not ask any controversial questions (e.g. challenging or difficult topics) and was directed to those clinicians who expressed willingness to engage in the CEN and was completely anonymous at all stages. The invitations to participate were sent through the BSPD membership with the authors of this papers having no access to participant contact information or any identifying information. No ethical approval was therefore required.

The survey consisted of 18 mixed format questions, including free text options, single answers and ranked options. These included demographics of the respondent's Paediatric Dentistry background, AI patients seen in their service and pathways including referral into specialist care and provisions for transitional care. Descriptive analysis was used to interpret the results and analysis was completed of the free-text data by a single-handed reviewer. The analysis was based on a recognised 6 stage format including familiarisation with the data, coding, identifying and reviewing of themes.<sup>11</sup> This method has been similarly used in other cross-sectional studies in the literature seeking views of the dental workforce.<sup>12</sup>

## Results

### Respondents

The survey was sent to 670 BSPD members, with a response rate of 17.2% (n=115). Respondents were from a mixture of all four UK home nations and were mainly based in the Hospital and Community Services as demonstrated in Figure 1. Most clinicians were based in England (72.6%, 82/113), followed by Scotland 13.3% (15/113), Wales 8.9% (10/113) and 5% (6/113) in Northern Ireland. The respondents were a mixture of consultants (28.7% n=33), specialists (24.4% n=28) and other dentists with an interest in Paediatric Dentistry through their BSPD membership (47% n=54). All 115 respondents answered the primary work role question and where the majority of their work was based. For the remaining questions there was a varied response rate as indicated in the text, with most questions answered by 93-99 clinicians.

**Figure 1: Respondents to survey primary work role**

### AI patients

The average number of new AI diagnoses seen per month was reported to be low. One to two patients diagnosed per month was the most common (48.5%, n=48/99), but many respondents would not see any new AI diagnoses (27.3%, n=27/99), with only 1 respondent reporting they would see more than 10 each month.

Current AI patients under care was also reported as low, with the majority of clinicians (81.8%, n=81/99) having between one and ten patients under their care. Figure 2 demonstrates the differences in responses to number of AI patients under care. Treatment appointments for AI were not reported as frequent with 84.7% (n=83/98) of respondents reporting less than five treatment appointments for AI patients per month. Another 12.2% (n=12/98) reported they carry out between five and ten appointments per month, with only 3.1% (n=3/98) carrying out more than 10 treatment appointments.

The patient age group most commonly seen was 6-12 years old (69.9%, n=65/93). The types of AI seen by respondents was varied with responses spread across all given definitions of: Hypomature, Hypoplastic, Hypocalcified, Hypoplasia/ hypomaturation/ taurodontism and mixed appearance.

**Figure 2: Bar-chart demonstrating how many AI patients are currently under care**

## Pathways

No clear AI referral pathway into specialist care was reported by 49.5% (n=47/95) of respondents, with another 19% (n=18/95) unsure if there was a pathway.

The majority of patients were referred into specialist care by their General Dental Practitioner (93.6%, n=88/94) with small numbers referred by their General Medical Practitioner (1.1%, n=1/94), self-referral (1.1%, n =1/94) or through other means (4.3%, n=4/94). A dedicated AI or anomaly clinic for paediatric patients was not common with 72.6% (n=69/95) of responses indicating there were no dedicated clinics, as can be seen in Figure 3.

**Figure 3: Bar-chart indicating whether there were dedicated AI or anomaly clinics.**

A clear transitional care pathway was deemed not to exist by 77.4% (n=72/93) of respondents, with only 22.6% (n=21/93) of respondents indicating there was. In terms of obstacles to providing a transition service, there were only 15 responses to this question. Of these 15 responses, 53.3% (n=8/15) did not think they were any obstacles.

Dedicated AI clinics for adult patients was not known to exist in most cases (56.8%, n=54/95) with another 35.8% (n=34/95) of respondents unsure if this was available.

Respondents were asked whether their patients transitioned to a number of different settings and selected all that applied. There was a combination of multi-disciplinary, general dental service and adult restorative transitions pathways utilised as indicated by 22 respondents (Figure 4). This was from a smaller number of respondents (19%). Specialist adult restorative services was the most popular transition pathway, followed by returning patients to primary care once treatment completed. Some respondents (n=11) indicated their patients would simply be discharged after a set period of care in their service. Interestingly, the yellow bar in Figure 4 demonstrates there were a number of respondents who were unsure if some of the transition pathways were available to them. In terms of justification for transition pathways, this was asked in a subsequent question, with 85.9% (n=73/85) of respondents thinking there is a need for a transition pathway for AI patients.

**Figure 4: Bar-chart indicating where the patients transition to following paediatric care**

## Free Text Analysis

The final four questions of the survey provided free text to answer the questions concerning transitional services, obstacles in providing these and any further comments around AI care. The data was analysed by the lead for the project. Five core themes were identified as shown in Figure 5 with examples of quotes related to the themes given.

The four questions included:

1. *Please expand on the above (e.g. what transition pathway is most common in your service?) (18 responses)*
2. *Are you facing any obstacles providing such a transition service? Please specify (15 responses)*
3. *Do you feel there is a need for a transition pathway? (85 responses)*
4. *Are there any other comments you wish to feed back around AI care? (37 responses)*

**Figure 5: Free text analysis themes**

## Discussion

This survey provides evidence towards the improvements required in paediatric pathways and transitional care arrangements for AI patients in the UK. For many clinicians the number of AI patients seen regularly is small. This therefore suggests the optimum management pathways and care decisions required are unfamiliar, and infrequently practiced. In general, it was found that no clear pathway into specialist care or a protocol for transitioning to adult services exists. The respondents strongly felt there is a need for a transition pathway with access to the appropriate specialist workforce highlighted as an area requiring improvement. The AI phenotype and patient wishes were deemed to be important considerations in the transition to adult care. A flexible and locally adaptable pathway was highlighted as essential.

The literature supports the uncertainty in providing the correct care for AI patients and the need for *lifelong* care.<sup>2,13</sup> Early diagnosis is important, with appropriate and timely multi-disciplinary care key.<sup>14</sup> The correct knowledge and skills in providing good quality care needs to be available to all, with access to specialists when needed. Anomalies associated with AI such as open bites, periodontal conditions, pulp stones and taurodontism all complicate care.<sup>15</sup> For provision of restorative care, indirect restorations have been shown to be superior to direct restorations for AI patients and should be provided as early as possible.<sup>16</sup> Long-term follow-up has shown that ceramic as a material choice can work well in young adults.<sup>17</sup> Careful treatment planning in the growing patient is required to execute indirect restorative work well. Combined with the challenges in providing the correct clinical care for AI patients, there is also known reservations surrounding newer aspects such as genetic testing for AI, with further exploration and education required.<sup>18</sup> The complexities of managing a CYP with AI can be well supported and addressed by access to the correct local specialist services.

Approximately half of the respondents were either specialists or consultants in Paediatric Dentistry (53%), with the remaining having an interest in Paediatric Dentistry. This allowed a well-rounded perspective from both specialists and non-specialist care providers and how they work together to provide care. The responses were from clinicians based throughout the UK which allowed a diverse range of opinions to be gathered with respondents able to express their views clearly through the 18 questions and further elaborate through the free-text comments in the final questions.

Limitations of this survey include that given it was voluntary, the respondents will naturally have been those with a special interest and investment in AI care for CYP. This self-selected group could therefore be a biased representation. The results however do raise valid points about difficulties accessing care and unclear pathways with useful information gathered in this context. There is a low response rate to this survey, however it was sent to a large number of clinicians associated with the speciality of Paediatric Dentistry. With the majority of respondents being either consultants or specialists in Paediatric Dentistry, the results are still important to consider, with these clinicians being responsible for their transfer from Paediatric to Adult services.

Other limitations include that not all questions were completed in full by every respondent, but all available data was reported; excluding questionnaires with incomplete answers would have reduced the volume of data available for analysis. Reasons why some of the data was missing / questions not answered by all clinicians, may have been that some respondents were not aware of the transition services available in their department/locality or were unsure of some of the answers. Furthermore, the small quantity of qualitative data



was analysed using thematic analysis by a single-handed reviewer; the authors felt this process was adequate given the free-text sections were limited.

It is clear there are regional and local variations in care and not one pathway or care model will be appropriate for every service. In the literature there is evidence of other areas of Paediatric Dental care pathways that require improvement and mirrors many of the issues found in this study.<sup>12</sup> Further explorations into local challenges faced by services would be beneficial. This should be alongside development of national protocols and guidelines to form a basis for service leads to refer to and mandate for certain types of care to be provided. Further research involving adult specialist service teams would also be beneficial with consensus reached on national referral criteria and acceptance for specialist care. In addition, discussion with clinicians in primary care along with the patients and families themselves will be crucial in developing robust transition pathways. Understanding the needs for children with AI is complicated with groups such the Clinical Excellence Network for AI patients, formed in 2019, being a method for strategic and multi-centred agendas to be discussed and researched. The overall aim of promoting clinical excellence for young patients with this condition ensures the importance of good AI care is recognised with this group's aim being to drive forward the required improvements nationally.

## **Conclusion**

Access to specialist paediatric care and transition to adult services is not readily available throughout the UK for AI patients. Clinicians in primary care who encounter this rare condition are therefore at risk of inadequate specialist support. There are several reasons for this identified, such as further education and awareness of AI throughout the dental workforce required. Access to specialist care and appropriate transition options for the individual patient is essential. There is a clear need to establish and improve existing care pathways and this should be approached nationally and adapted locally.

## **Author Contributions**

SA, SP and FS approved the survey questions and reviewed the results and manuscript. FL led the survey roll out, analysis and manuscript writing.

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