Rehabilitation of Children with Ectodermal Dysplasia. Part 1: An International Delphi Study

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Purpose: *An international Delphi study was undertaken to determine by consensus an agreed approach to the management of children with dental manifestations of ectodermal dysplasia, including the use of dental implants. This was done using a questionnaire developed by an interdisciplinary team.* ***Materials and Methods:*** *The Delphi study questionnaire was built around 19 areas of clinical relevance and included 90 items. Topic areas included dental disability; initial diagnosis; global disability; oral health aspects of dental treatment (orthodontics, hypodontia, anodontia, implants); and case studies of selected treatment options. Eleven teams from six countries contributed to three iterations of the questionnaire. An algorithm was designed to standardize analysis of the questionnaire answers, all of which were blinded to ensure anonymity. The second and third rounds of the questionnaire excluded previously agreed-upon items but included the responses to the questions from the earlier rounds. The nonconsensus items inquired about the use of radiographs at initial diagnosis; sedation of an uncooperative child; use of a pretreatment questionnaire; the age range for specific treatments (eg, dentures, orthodontics, implants); specific uses of implants (eg, partial prostheses, overdentures, cantilevered prostheses); and case study 2. The residual nonconsensus questions were subsequently discussed at a 2-day meeting.* ***Results:*** *Among the 90 questions and partial questions, there was progressive consensus, with agreements in rounds 1, 2, and 3 of 61%, 21%, and 8%, respectively. At the conclusion of round 3, there was 90% agreement and it was considered that the nonconsensus items required in-depth face-to-face discussion at a consensus meeting, which is described in part 2 of the study.* ***Conclusion:*** *The Delphi study provided an opportunity to engage specialist teams in recognized centers to integrate their clinical knowledge and draw on published data to develop a consensus of evidence-based responses.* Int J Oral Maxillofac Implants 2013;28:1090–1100. doi: 10.11607/jomi.2980

Key words: *consensus, Delphi study, ectodermal dysplasia*

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The treatment of dental problems related to ectodermal dysplasia (ED) is challenging and best managed by multidisciplinary teams,[1] the combined experience and knowledge of which could potentially enhance care in this field. Following informal discussions, it was agreed that an international Delphi study should be undertaken to establish a consensus on key treatment decisions using a specific questionnaire designed to address the topic. Given the widely held views on the benefits of dental implant treatment in this patient group, significant emphasis was placed on this aspect of care.

It was recognized that other study designs were possible; however, the Delphi technique was selected for this complex clinical area to allow interaction across many specialist groups with extensive experience in the field to ensure that the results would reflect an agreed-upon international perspective. No other study design would provide ongoing interaction to reach consensus. It was also planned that, if there was a lack of consensus after an appropriate number of iterations, a consensus meeting would be held.

The complexity of presentation of such cases is acknowledged,[2] and while the predictability of oral implants in adult patient care is recognized, there are reservations about the use of implants in the growing bone of children, particularly with regard to timing.[3,4] Patients require detailed diagnostic and treatment decisions; however, the lack of alveolar bone associated with congenitally missing teeth can severely compromise ideal implant placement. Current data recommend that the placement of implants in adolescent patients be delayed until 13 to 16 years of age.[5] The specific requirements for implant management in children with ED with anodontia and hypodontia represent a new and challenging frontier of oral rehabilitation.[6]

Growing patients, particularly those presenting with ED, often require prosthodontic rehabilitation for the restoration of edentulous areas resulting from congenital anodontia and hypodontia. Historically, these have been treated with removable prostheses prior to skeletal and dental maturation. This technique continues to play a role in rehabilitation, and children usually adapt well to their prostheses. Nevertheless, increased alveolar bone resorption in partially dentate individuals results in increased periodontal complications, and increased dental caries are more common with the use of removable partial dentures (RPDs).

DENTAL IMPLANTS IN CHILDREN AND ADOLESCENTS

Over the last 20 years, with confirmation of the predictability of endosseous implants, there has been increasing interest in using this treatment in growing patients for prosthodontic rehabilitation. However, this creates additional concerns that are not seen in the adult. Dental and skeletal growth are of concern in the predictability of implant success because growing bone represents a confounding variable for implants.[7] Hypodontia and anodontia, as seen in ED, are associated with dental deficits and facial changes, which have social consequences, especially in early adolescence.[8]

Conservation of bone may be the most important reason for the use of dental implants in growing patients.[3,4,9] It is important to also note that, in children with congenital anodontia or hypodontia, little or no alveolar bone is present.

Dental implant treatment in children is a relatively new treatment modality, and the impact that bone-supported prostheses might have on facial growth, or conversely, the influence of growth on the longevity of implant prostheses, is unclear. There are two primary concerns:

1. If implants are present during facial growth, how do their physical relationships change relative to each other and to the external contours of the bone? Do they become embedded, relocated, or displaced? These outcomes are possible, as implants (in contrast to teeth) do not have compensatory eruption or other physiologic movement.
2. What is the effect of an implant prosthesis on growth? Do fixed prostheses attached to implants in a growth area inhibit growth, and what superstructure design might therefore be required to compensate for this?

Craniofacial growth and dental development are complex and have been extensively studied and documented, but there is limited information on the use of implants in growing bones. The effects that implants might have on growth and development can be extrapolated from the considerable information available on ankylosed primary teeth and traumatized permanent teeth.

Data are available to document the large amount of growth that occurs in the maxillary posterior segments and thus the potential for osseointegrated implants to become buried in alveolar bone.[10] Growth disturbances frequently accompany ankylosed teeth because of the adaptive and eruptive changes that occur when vertical, lateral, anterior, and posterior growth are affected. The growth process involves new alveolar bone formation, whereas ankylosis is accompanied by an arrest of eruption and alveolar bone growth in the affected area. An osseointegrated implant may behave similarly, with the same lack of alveolar growth and eruption.[11] This has been reported in animal models[12] and with the use of implants in growing patients with ED.[7]

Studies by Bjork and Skieller[13] and Bjork[14] used small pin implants in bone to study growth and bony change to increase understanding of the behavior of implants during growth. Bjork implanted 0.5- × 1.5-mm tantalum pins in the jaws of children to serve as landmarks for longitudinal cephalometric studies. Although the majority of implants were stable, pins affected by growth were not. Pins placed in the path of erupting teeth and those placed in bone that was undergoing resorption became displaced. Orthodontic tooth movement also displaced pins: Nearly all pins placed in resorptive areas, such as the anterior mandibular ramus or anterior maxilla, were lost and required replacement. Pins placed in areas of appositional bone growth gradually became embedded.

The cause of hypodontia, the gender of the patient, and skeletal maturity are key factors in the assessment of the timing of implant placement. Although a congenitally missing maxillary lateral incisor is an excellent site for implant placement, it is acknowledged[15] that this apparently simple restoration may be associated with complications. In a review of 42 implants placed in 34 patients with a mean age of 15 years, Lederman and coworkers[15] reported a 90% success rate with a mean follow-up period (postloading) of 35.5 months. This study reported positive soft and osseous tissue reactions to the implants and found that the majority of failures occurred because of subsequent traumatic injuries sustained during the healing phase after implant placement. The most common postloading complication was the failure of the implant to respond to vertical growth of the adjacent teeth and alveolar bone, caused by its ankylotic nature.

There is controversy concerning the timing of placement of dental implants in young children, as there is little published material on the technique and its long-term consequences. It is generally believed that implants act similar to infraoccluded ankylosed teeth and do not move with growing bone.[11] Recent animal research has confirmed that most implants become osseointegrated in growing jaws; however, there is no evidence that implants behave like teeth during development.

In the mandible, implants may become displaced lingually, while in the maxilla they may become displaced palatally and superiorly and may not follow the usual downward and forward growth of this bone.[16] This is important when considering the placement of implants in the anterior maxilla. Further, implants appear to retard alveolar growth locally and change the eruptive paths of distally positioned tooth buds.

Implant restoration has, in general, not been considered in children before completion of bone growth. It should be noted, however, that in children with conditions such as ED, alveolar bone does not develop where teeth are congenitally absent. Consequently, it may be possible to place implants much earlier in these children than in those with alveolar bone. Several papers have reported the successful use of implants in growing children.[8,17–21] A consensus conference recommended the placement of implants in the canine region of the anodontic mandible at 6 to 8 years of age.[22] Guckes et al[20] reported on the placement of 264 implants in 51 patients with ED. A higher success rate was seen in the mandible (91%) compared with the maxilla (76%), with the predominant location of failures being the anterior maxilla. Sweeney et al[23] identified increased failure rates for implants placed in areas with a lack of bone volume.

The current questions and difficulties in the management of children with ED include the need for early intervention to restore function and esthetics to assist psychosocial development. As a result, a number of questions need to be addressed:

- At what age should prostheses be placed?
- What are the most appropriate designs for removable appliances in young children?
- If removable appliances prove inadequate, when is the most appropriate time to place implants?
- In which areas of the jaws should implants be placed?
- Does the early placement of implants interfere with growth and development?
- Management requires multidisciplinary planning; can standard treatment plans be developed for patients with anodontia and different patterns of hypodontia?

Consensus reports have been published on aspects of patient management. Relevant data were summarized by Koch et al[22] and Bergendal et al[24] and include the following.

1. The available knowledge suggests that the use of implants in healthy children is possible.
2. According to present knowledge, no health hazards or contraindications of implants in young patients have been reported. In addition to generally accepted contraindications for oral implant treatment, there may be others, including rare conditions that affect bone metabolism.
3. There is no fixed age for implant placement. Instead, biologic age should be determined regarding growth and skeletal development, which should be complete or nearly complete, as assessed by longitudinal body height measurement and hand-wrist radiographs.[22] In cases of anodontia and severe oligodontia, however, oral implants may be placed before the pubertal growth spurt.

4. In considering the advantages and disadvantages of placing oral implants in growing individuals, a number of concerns have been noted, including dentoalveolar development, craniofacial growth, short- and long-term perspectives, and function and esthetics. The advantages of implants for replacing single and multiple missing teeth include the following: *(1)* patients receive a fixed prosthesis to improve function and esthetics without involving adjacent teeth; *(2)* in cases of tooth loss from trauma and general spacing, placement of implants is an alternative; *(3)* in cases of multiple missing teeth, the therapeutic alternatives are fewer, and placement of oral implants may allow good oral function to be established and alveolar bone to be maintained. The disadvantages of implants in young people include the fact that implants do not follow the growth of alveolar bone and the risk of infraocclusion of implant-retained prostheses, which may negatively affect function, periodontal health, and esthetics.
5. In cases of anodontia, placement of implants ensures a stable prosthesis with improved function, which leads to reduced psychosocial stress. There are no known disadvantages compared with other prosthetic treatments,[25] although esthetics may be achieved equally well with removable prostheses.
6. A multidisciplinary team approach is recommended and is mandatory for treatment of patients with extensive lack of teeth, anodontia, or rare syndromes. Early diagnosis is advocated and alternative treatment modalities must always be considered.[1,26,27]
7. Ethical aspects on the use of oral implants in young individuals have been addressed as follows: *(1)* Treatment should be performed in the best interests of the child, per the United Nations Convention on the Rights of the Child; *(2)* treatment should be preceded by agreement following discussion and approval from the patient and/or the caregivers; *(3)* treatment should be performed by specially trained and experienced professionals; and *(4)* treatment should be documented by long-term follow-up.[5]
8. An oral care program for individuals with ED should incorporate[24] a multidisciplinary team, preferably including pediatric dentistry, orthodontics, prosthodontics, and maxillofacial surgery, and the patient/family should be involved in treatment planning.
9. While there is considerable international activity on the multidisciplinary treatment of hypodontia and a widespread recognition of the value of implant treatment, there is currently no international consensus on key treatment decisions, including, crucially, the appropriateness of implant placement in the younger patient.

MATERIALS AND METHODS

A questionnaire covering 19 areas of particular clinical relevance was developed in consultation with clinical specialists in prosthodontics, orthodontics, and pediatric dentistry at Westmead Centre for Oral Health, Sydney, Australia, to determine a clinical protocol to manage ED with hypodontia. Validation of the questionnaire was provided through consistent responses from a pilot trial with members of the Sydney team.

The breadth of questions selected was designed to address key areas of interest in clinical decision making. It was recognized that, for some questions, sufficient published data were available; however, the questions were included to ensure that the issues were addressed comprehensively. In addition, three case histories (one patient was presented at two different times of life) and images were included to focus considerations and comments on the common clinical presentations of the syndrome. These clinical cases are part of the database of patients at the Westmead Centre for Oral Health.

The clinical decisions and agreements reached were considered to be of significance in case management.

The questionnaire consisted of topic areas covering:

- Dental disability (1 item)
- Initial diagnosis (8 items or partial items)
- Global disability (10 items or partial items)
- Oral health (8 items or partial items)
- Aspects of dental treatment: orthodontic (4 items or partial items), hypodontia (10 items or partial items), anodontia (6 items or partial items), implants (24 items or partial items)
- Four case studies involving: *(1)* a 3-year-old (5 items or partial items), *(2)* a 15- to 19-year-old (6 items or partial items), *(3)* another 3-year-old (4 items or partial items), and *(4)* a 5-year-old (4 items or partial items)

Ninety items or partial items were presented for completion. Also included with each question was a request for comment on the relevance to the topic when considering the validity of the items presented. Forty-nine of the clinically specific items, or partial items, have two parts for responses. The first part was a 5-point scale on which the participants could indicate their level of consensus, and the second part invited an opinion on that statement. The other items in the questionnaire invited opinion only.

An international panel of expert clinicians in pediatric dentistry, prosthodontics, and orthodontics was invited to be part of a study based on a consensus of ideas. Each expert team within their global location was deemed to represent that location. Acceptance of participation required the signing of a letter of agreement for the Sydney team.

Table 1 The Delphi Study Process

At each center	Coordination in Sydney
• Acceptance of study participation by completion and return of letter of commitment	• Receipt and de-identification of questionnaire responses by the study coordinator
• Receipt by email of the questionnaire by the team coordinator	• Collation of responses to individual items
• Meeting of colleagues in each study group to consider questionnaire items and provide appropriate responses based on available evidence and/or expert opinion	• Analyses of the items received from each center
• Complete questionnaire online and return by email to the coordinating group in Sydney	• Construction of the following round of items based on criteria
	• Resending of the revised questionnaire to each center by email
	• Review of progress after the third iteration
	• Possible reconsideration of the process for further revisions and iterations
	• Preparation of a progress report

Table 2 Summary of the Proposed Timeline of the Study Iteration

Round	Receive questionnaire, complete, and return to Sydney	Revise, consider in detail all responses to the questionnaire in Sydney, and return to centers	Approximate times
1	2 months	2 months	4 months
2	2 months	1 months	3 months
3+	2 months	1 months	3 months
Total time			10 months

At consensus meeting:
- Review and discuss Delphi study outcomes and consider the nonconsensus items
- Discuss the plan for a pilot study
- Plan requirements and participation of expert groups for an international multicenter clinical study of implant rehabilitation of ED anodontia and hypodontia in children 8 to 15 years of age

Expert panels from six countries participated and represented the individual teams.

Previous Delphi studies have reported that, generally, three iterations are needed to provide a convergence of ideas.[28] Each round or iteration of the questionnaire provided developing consensus on the questions proposed and required the collation of responses to the items from each center. The responses to questions allowed removal of those questions for which there was consensus and the resending of a revised questionnaire for further consideration.

After each iteration, the questions that remained required formatting of the questionnaire in the same manner to allow comparisons across the iterations as well as to reduce bias. Any item that needed to be altered for clarification was reformatted by the Sydney team following suggestions from participating group leaders.

The requirement for consensus or nonconsensus at each round or iteration was repeated and the process was continued to reach increasing levels of agreement. It was intended that, if after a maximum of five and possibly three rounds,[28] there remained items on which no consensus had been reached, the remaining matters would be discussed at a consensus meeting.[29]

The Delphi study process is summarized in Table 1. It was anticipated that the timing of the process of sending, returning, revision, and resending of the questionnaire would take 3 to 4 months for each iteration.

Analysis of Delphi Study Questionnaire

At each round and questionnaire iteration, agreement was determined by the use of a progressive algorithm developed for the purpose by the Sydney team. The algorithm was used to determine whether agreement was achieved for each individual question in each round. All written responses and sums of responses received from individual teams for which there was no consensus were used to redraft each specific question. This ensured that the following iteration of the questions for the next round clarified the wording of the questions that had been raised by the teams. The proposed progressive algorithm prescribed three combinations of responses to allow agreement:

- Responses with only "agree" + "definitely agree"
- Responses with only "don't know" + "agree" + "definitely agree"
- Responses with a maximum of two "not agree" + "agree" + "definitely agree"

With each set of answers, written responses were also received. All comments on questions were scrutinized by two members of the Sydney team, who determined independently whether the comments contributed to a level of agreement or nonagreement and whether there was a need to redraft specific questions for the next iteration of the questionnaire. If required, this information was further discussed with members of the Sydney team to clarify agreement. For there to be consensus, similar responses to questions had to be reported by at least 75% of the expert groups.

A proposed timeline is summarized in Table 2.

RESULTS

The three rounds of the Delphi study yielded progressive agreement. Round 1 yielded consensus for 57 questions and partial questions, with 38 nonconsensus items remaining; round 2 yielded consensus on a further 24 questions and partial questions, with 16 nonconsensus items remaining; and round 3 yielded consensus on a further 7 questions, leaving a residual 8 nonconsensus items. The Delphi sequence resulted in consensus on 88 questions or partial questions at the completion of the third and final round.

The residual nonconsensus items were to be considered at a consensus meeting to allow face-to-face discussion with key members of each team. Table 3 tabulates the consensus agreements with the three rounds and indicates progressive consensus (C) over the three rounds and the progressive reduction of nonconsensus items (N). Figure 1 clearly displays the percentage of progressive consensus, beginning with 61% in round 1, an additional 21% in round 2, and an additional 8% in round 3, with a residual 10% of nonconsensus items.

Figure 2 provides the details of each question and summarizes the round in which consensus was reached, as well as the specifics of the algorithm that defined the level of agreement or disagreement for each question. This information relates to specific questions and partial questions #1 to #15, each of which concern aspects of assessment that would be considered in the management of ED patients. Questions and partial questions for #16 to #19 are specific to clinical cases that are representative of those present for assessment and management.

Table 3 Overview of Responses for the Three Rounds

Question	Round 1	Round 2	Round 3	Question	Round 1	Round 2	Round 3
1	C			12a_3	C		
2	C			12a_4	C		
3a	N	N	N	12a_5	C		
3b	N	C		12a_6	C		
3c	N	C		13a	N	N	N
3d	N	C		13b_1	C		
4a	C			13b_2	C		
4b	C			13c	C		
4c	C			13d	C		
4d	C			13e_1	N	C	
4e	N	C		13e_2	N	C	
4f	N	N	C	13f	N	C	
5a	C			13g_1	N	C	
5b(i)	N			13g_2	N	C	
5b(ii)	N			13h_1	C		
5b_1(i)		N	N	13h_2	C		
5b_1(ii)		C		13i_1	N	N	N
5b_2(i)		C		13i_2	N	N	C
5b_2(ii)		N	C	13j_1	N	N	C
6a	C			13j_2	N	N	C
6b	N	N	C	13k_1	N	N	N
7a	N	N	N	13k_2	N	C	
7b	C			13k_3	N	N	C
7c	C			14a	C		
8a	N	C		14b	C		
8b	C			14c(i)	N	C	
9a	C			14c(ii)	C		
9b	C			14c(iii)	C		
9c	C			15a	N	C	
9d	C			15b	N	C	
9e	C			16a	N	C	
9f	C			16b	C		
9g	C			16c	C		
9h	C			16d	N	C	
10a	C			16e	N	C	
10b	C			17a	C		
10c	C			17b	C		
10d	C			17c	C		
11a_1	C			17d	N	N	N/C
11a_2	N	N	N	17e	C		
11a_3	N	N	N	17f	C		
11a_4	C			18a	N	C	
11a_5	C			18b	C		
11a_6	C			18c	N	C	
11b_1	C			18d	N	C	
11b_2	N	C		19a	C		
11b_3	C			19B	C		
11b_4	C			19c	C		
12a_1	C			19d	N	C	
12a_2	C						

C = consensus; N = no consensus.

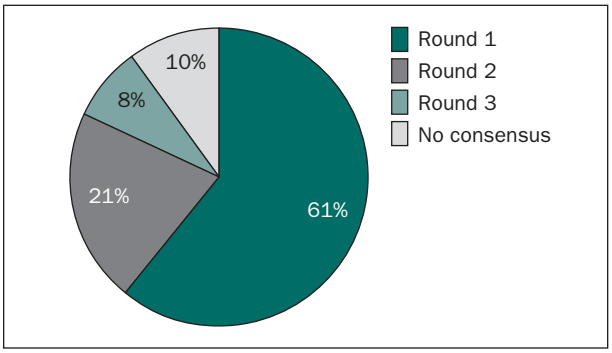

Fig 1 Results: Percentage consensus on all questions by round.

DISCUSSION

A Delphi study method was selected for this complex clinical area to allow interaction across a total of 11 specialist centers. The centers were selected because they were recognized for their expertise in ED case management, and each team agreed to provide considered responses to questions regarding clinical management. The Sydney team considered this to be the optimal mechanism to resolve matters of clinical concern in the diagnosis and treatment of ED cases. This approach had the advantage of an ongoing engagement of international team members with answers to specific questions and qualifying written responses to explain the decisions. This process also had the advantage that it would clearly reflect an international perspective and lead to a consensus meeting if necessary. The Sydney team regarded the Delphi study outcomes to be the first step in developing a multicenter clinical trial.

An important aspect of this Delphi study design was the request for specific comments from the centers on each question. This information ensured that, for successive rounds, the questions were revised to incorporate feedback from each center to help address matters of clarity. These modifications to the wording of questions facilitated progressive agreement. However, it was recognized that after three rounds, with agreement reached for the majority of questions, further consideration would require face-to-face discussion through a consensus meeting.

Table 3 tabulates the consensus or nonconsensus achieved in each of the three rounds. Figure 2 provides the details of each question and indicates whether consensus was reached or not.

Round 1 resulted in 61% agreement. This involved the following questions: 1 (teeth absent in ED for which there was data), 2 (need for genetic counseling), 4 (retention of primary teeth), 5a (need to address a patient's specific concerns), 6 (psychologic implications), 7b and 7c (function and speech), 8b (nutrition in relation to oral and general health), 9 (caries risk and home care), 10 (need for orthodontic treatment), 11 in part (age for treatment of hypodontia), 12 in part (age for treatment of anodontia), 13 in part (implants in growing bone), and 14 in part (evaluation of implant treatment). Case studies were described in questions 16, 17, 18, and 19, and there was agreement for part of each question.

It was recognized that, although the cases were of a commonly presenting form, they nevertheless presented some difficulty for centers, as the case descriptions and radiographic information were limited. However, the essential features to be considered were presented. The information requested was a consolidation of the general questions.

Round 2 resulted in an additional 21% agreement, which involved the following questions: 3 (when to use radiographs), 4e (retention of ankylosed primary teeth), 5b_1(ii) (need for sedation to restore anterior teeth), 5b_2(i) (need for sedation to take an impression), 8a (need for pretreatment questionnaire on nutrition), 11b_2 (need for retention in the use of a RPD), 13e_1 (effects of implants in maxilla), 13e_2 (effects of implants in mandible), 13f (effect of implant on craniofacial bone), 13g_1 (effect of bone growth on implant position in maxilla), 13g_2 (effect of bone growth on implant position in mandible), 13k_2 (an implant overdenture in growing bone), 14c(i) (follow-up radiograph/panoramic), 15 (case 1), 16a (treatment plan), 16d (age for treatment to begin), 16e (pharmacological management if required), 18a (age to begin treatment of mandible), 18c (management of a maxillary anterior diastema and why), 18d (orthodontics or restorative option), and 19d (retention of bone in the posterior maxilla).

Round 3 resulted in additional agreement for 8% of the questions, which involved the following: 4f (retention of ankylosed primary teeth inhibiting alveolar growth), 5b_2(ii) (use of general anesthesia to restore anterior teeth), 6b (pretreatment assessment through questionnaires), 13i_2 (implant spacing for three-unit implant denture or implant cantilever denture in mandible), 13j_1 (implant spacing for three-unit implant denture or implant cantilever denture in maxilla), 13j_2 (three-unit implant denture or two-implant cantilever denture in growing bone), and 13k_3 (the form of attachment to be used with an implant overdenture).

There was a 10% residual of nonconsensus items after the third round of questionnaire. These questions were considered at a consensus meeting.

The Delphi study iterations continued for significantly longer than the 10 months (Table 2) originally envisaged; the duration varied with the time available for each group to meet, to consider the questions, and return their individual responses. In addition, more time was required by the Sydney team to consider the de-identified answers and feedback from each of the 10 teams and to integrate the feedback where required into revised questions.

Q1.	**Do you agree that the teeth most frequently absent (excluding third molars) are?**	
	(a) Maxillary lateral incisors,	
	(b) Second premolars	
	(c) Mandibular central incisors	
	(d) First premolars	
	(e) Second molars	
	(f) Canines	
	(g) First molars	
	(h) Maxillary central incisors	AGREE
Q2.	**Do you agree that genetic counseling is a necessity?**	DEFINITELY AGREE
Q3.	**Radiographs: At what age—(a) 3 to 5 years, (b) 6 to 8 years, (c) 9+ years—should each type of radiograph be taken?**	
	(a) OPG	NO CONSENSUS
	(b) Periapicals	9+ YEARS
	(c) Bitewing	6 TO 8 YEARS
	(d) Lateral cephalometric	9+ YEARS
Q4.	**Retention of primary teeth: Please comment on (a) to (d).**	
	(b) Primary teeth should be retained into adolescence if root form is present and the teeth are stable.	DEFINITELY AGREE
	(c) Primary tooth size discrepancy needs to be considered in initial treatment.	AGREE
	(d) Which primary teeth should be kept?	CONDITIONAL
	(e) Should primary teeth be retained if they become ankylosed?	AGREE
	(f) Does the retention of ankylosed primary teeth (where there is no permanent successor) inhibit alveolar bone growth?	DON'T KNOW
Q5.	**Do you agree with the following statements?**	
	(a) Specific personal concerns of each patient need to be considered and addressed.	DEFINITELY AGREE
	(b) Parents often have legitimate but unrealistic demands in relation to the provision of treatment for their child. Is it acceptable to sedate/manage under general anesthesia a precooperative child to provide treatment for hypodontia in the following situations?	
	1. Is it acceptable to sedate a precooperative child to provide treatment for hypodontia to:	
	(i) Take an impression	NO CONSENSUS
	(ii) Restore anterior teeth	AGREE
	2. Is it acceptable to manage under general anesthesia a precooperative child to provide treatment for hypodontia to:	
	(i) Take an impression	DEFINITELY NOT AGREE
	(ii) Restore anterior teeth	AGREE
Q6.	**Psychologic implications need to be considered as a patient-specific problem.**	
	(a) It is essential to consider the following: behavior (concentration, irritability); well-being (nervous, lonely, easily upset, cheerful); self-esteem (school, sport, friendships, appearance, family relationships).	DEFINITELY AGREE
	(b) A pretreatment assessment is required by a specialist in pediatric dentistry who has "child competency" and/or by a clinical psychologist, through questionnaires, including oral health–related quality of life measures completed by: parent(s) and child.	NOT AGREE
Q7.	**Orofacial function needs to be considered as a patient-specific problem and the assistance of parents is required.**	
	(a) A pretreatment assessment questionnaire is required to be completed by: parent(s) and child.	NO CONSENSUS
	(b) Orofacial function is important in relation to oral and general health, including:	
	• Jaw movements (open-close, lateral, protrusive)	
	• Tongue movements	
	• Speech	DON'T KNOW
	(c) Speech assessment may also be required by a speech pathologist.	DEFINITELY AGREE
Q8.	**Nutrition is a patient-specific problem and needs to be considered by a dietitian.**	
	(a) A pretreatment assessment questionnaire is required to be completed by: parent(s) and child.	NOT AGREE
	(b) This is important in relation to oral health and general health and is age specific:	
	• The parent's knowledge of diet and dental implications is crucial	
	• The age of introduction of refined foods is important	
	• The source of food and nutrition: parent, grandparent, or other relative	AGREE
Q9.	**Caries risk and home care need to be considered.**	
	(a) Patient dental knowledge of oral hygiene status must be discussed.	DEFINITELY AGREE
	(b) Has dental advice been given by dentist, health clinic, relative, friend?	DEFINITELY AGREE
	(c) Medical factors and oral health need to be considered.	DEFINITELY AGREE
	(d) Access to dental services needs to be discussed.	DEFINITELY AGREE
	(e) Dental caries risk should be assessed.	DEFINITELY AGREE
	(f) Dental caries detection is helpful.	DEFINITELY AGREE
	(g) Regular use of toothbrush and fluoride toothpaste is required.	DEFINITELY AGREE
	(h) Saliva tests should be performed (volume, flow rate, quality).	AGREE

Fig 2 Summary of survey questions and consensus reached.

Q10. Orthodontic treatment	
(a) Interceptive orthodontics may be needed to control a deep anterior overbite.	AGREE
(b) Distribution of edentulous spaces should be managed to improve prosthetic treatment, esthetics, and possible implant placement.	DEFINITELY AGREE
(c) Dentoalveolar and jaw growth modification (such as with functional appliances) may be needed to improve facial proportions.	DEFINITELY AGREE
(d) Provisional implants should be used to provide orthodontic anchorage.	AGREE
Q11. Hypodontia	
(a) The following suggests an age range for specific treatment possibilities.	
1. 0 to 2 years: No treatment	AGREE
2. 2 to 3 years: RPD	NO CONSENSUS
3. 6 to 10 years: orthodontics/RPD with implants	NO CONSENSUS
4. 10 to 14 years: orthodontics/RPD/implant-fixed denture	AGREE
5. 14 to 18 years: orthodontics/RPD/implant-fixed denture	AGREE
6. > 18 years: RPD or implant fixed denture	DEFINITELY AGREE
(b) Factors in the use of removable partial dentures in young children:	
1. Tolerance	AGREE
2. Retention	AGREE
3. Use of overdentures	DEFINITELY AGREE
4. Maintenance or adjustment of vertical dimension	DEFINITELY AGREE
Q12. Anodontia: The following suggests age ranges for specific treatment possibilities:	
1. 0 to 2 years: No treatment	DEFINITELY AGREE
2. 2 to 6 years: Maxillary complete RDP	AGREE
3. 6 to 10 years: Maxillary complete RDP or implants	DEFINITELY AGREE
4. 10 to 14 years: Maxillary complete RDP or implant overdenture	DEFINITELY AGREE
5. 14 to 18 years: maxillary and mandibular complete RDP or implant overdenture or fixed prosthesis and implants	AGREE
6. > 18 years: maxillary and mandibular complete RDP or implant overdenture or fixed prosthesis and implants	DEFINITELY AGREE
Q13. Implants in growing bone (in the absence of alveolar bone) In considering the questions below, please indicate how they may apply to each age group, based on clinical experience and outcome data published in the scientific literature. Please consider the placement of implants in children with severe hypodontia, as opposed to those children who may have suffered tooth loss and normal alveolar bone is present.	
(a) Are there particular ages at which implants may be successfully placed in children?	NO CONSENSUS
(b) In which areas of the jaws are implants more predictable?	
1. Maxilla: anterior/posterior	ANTERIOR
2. Mandible: anterior/posterior	ANTERIOR
(c) What is the effect of placing implants in the maxillary tuberosity prior to pneumatization of the sinus?	DON'T KNOW
(d) At what age should the placement of implants be considered in the mandibular canine region?	5 TO 10 YEARS
(e) What are the effects of implants on growing alveolar bone?	
1. Maxilla: anterior/posterior	INHIBITS GROWTH
2. Mandible: anterior/posterior	INHIBITS GROWTH
(f) What are the effects of implants on growing craniofacial bone?	NO EFFECT
(g) What are the effects of bone growth on implant position?	
1. Maxilla: anterior/posterior	SUBMERGE/MALPOSITION
2. Mandible: anterior/posterior	SUBMERGE/MALPOSITION
(h) Can implants be placed adjacent to natural teeth in growing bone?	
1. Maxilla: anterior/posterior	NO
2. Mandible: anterior/posterior	NO
(i) Should implants be spaced, as for a three-unit RDP or two-unit cantilever denture, or placed adjacent to each other, as for separate crowns?	
1. Maxilla: anterior/posterior	NO CONSENSUS
2. Mandible: anterior/posterior	RDP
(j) Is an implant-supported three-unit denture or a two-unit cantilever denture more appropriate in growing bone?	
1. Maxilla: anterior/posterior	NO IMPLANTS
2. Mandible: anterior/posterior	NEITHER
(k) Is an implant overdenture more appropriate in growing bone?	
1. Maxilla: anterior/posterior	NO CONSENSUS
2. Mandible: anterior/posterior	YES
3. What form of precision attachment is desirable (eg, bar)?	BALL
Q14. Evaluation of implant treatment	
(a) Use of standardized radiographs is crucial for comparative measurements of bone levels at follow-up assessments.	DEFINITELY AGREE
(b) Follow-up assessment is required annually, or every 2 years.	AGREE
(c) Which radiographs should be used for follow-up?	
i. OPG	YES
ii. Cephalometrics	NO
iii. Periapicals	YES

Fig 2 continued Summary of survey questions and consensus reached.

Q15. Surgical technique for implant placement	
(a) A two-stage surgery is preferred.	AGREE
(b) A single-stage surgery is preferred.	AGREE
CASE STUDY 1	
Q16. Consider the following case: A 3-year-old female with sex-linked ectodermal dysplasia. What might be:	
(a) Your treatment plan for this child	COMPOSITES
(b) The restoration of the anterior teeth	COMPOSITES
(c) The plan for retention of the primary molars	RETAIN
(d) The ideal age at which treatment might commence	3 YEARS
(e) The plan for use of pharmacological behavior management techniques for a precooperative child	YES
CASE STUDY 2	
Q17. Consider treatment for the same patient at 15 years and 19 years. What might be:	
(a) The plan for retention of the primary maxillary canines	YES
(b) The plan for retention of the primary molars	YES
(c) The time at which these teeth may be removed	START OF TREATMENT
(d) The treatment for loss of vertical dimension associated with primary molar ankylosis	
At 15 years:	NO CONSENSUS
At 19 years:	IMPLANT
(e) The importance of the mandibular right second permanent molar in the overall treatment plan	RETAIN AND USE
(f) The possible prosthodontic management of the maxilla considering the amount of bone in the tuberosity.	BONE GRAFT
CASE STUDY 3	
Q18. Consider the following case of a 3-year-old child.	
(a) At what age would you consider treating the mandible and why?	3 TO 5 YEARS
(b) What are the possible options for provision of teeth in the mandible?	RPD
(c) How would you manage the maxillary anterior diastema?	NO TREATMENT
(d) Is it preferable to close diastema orthodontically or to attempt to build up the mesial of the central incisor?	ORTHODONTICS
CASE STUDY 4	
Q19. Consider the following case of a 5-year-old child.	
(a) At what age would you consider treating the mandible and why?	5 YEARS
(b) What are the possible options for provision of teeth in the mandible?	IMPLANT AND OVERDENTURE
(c) What are the possible options for provision of teeth in the maxilla?	RPD
(d) How might bone be maintained in the posterior maxilla?	NO TREATMENT

Fig 2 continued Summary of survey questions and consensus reached.

CONCLUSION

The Delphi study method proved to be successful in addressing complex management questions for which there was uncertainty and disagreement among clinicians managing ectodermal dysplasia (ED) patients with varying tooth and alveolar bone loss. The need for such an approach arose because of the uncertainty by the interdisciplinary Sydney team in managing such cases at the Westmead Centre for Oral Health and was catalyzed by the regular and predictable use of implants in adult oral rehabilitation. This was especially the case with oral rehabilitation treatment planning and outcome expectations of young ED cases, in whom bone growth continued but where there was an increasing requirement to manage the functional, esthetic, and psychosocial needs of these patients.

The study required that a specific protocol be followed by the coordinating team as a crucial component of study design to ensure validity. The defined study design also provided the opportunity for others to repeat the study.

Development of the questionnaire required time to determine: *(1)* the topic areas, *(2)* individual questions to unambiguously address important case assessment and treatment planning issues, and *(3)* and the inclusion of case histories with specific questions that required careful consideration by individual teams. The Sydney team believed that the extensive nature of the questionnaire (90 questions or partial questions) was needed to comprehensively address these requirements.

The other crucial matter was for the individual teams to agree to the protocol and for each team to meet to consider the questionnaire and to provide written

feedback to the Sydney team in addition to addressing the specific questions. There were uncertainties in the wording of some questions, which were addressed in the following iteration. In this way, specific questions were rewritten based on feedback to address the issues raised and ensure clarity. Although there were 11 international teams, each with long-standing experience in managing ED patients, there were considerable differences in clinical approaches, which contributed to the need for ongoing clarification with each iteration. What was remarkable was the degree of consensus reached initially and progressively with the following two rounds. However, it was apparent that after three rounds, there was a need for a face-to-face meeting to consider the remaining questions (summarized in Fig 2).

ACKNOWLEDGMENTS

The Sydney team wishes to acknowledge the commitment of each of the participating teams, their experience in management of ED rehabilitation, and their helpful and frank commentary on questions that required clarification, specifically those questions requiring a second and third round. This encouraging commitment to international cooperation is an example of the benefit of sharing of knowledge for the ultimate benefit of patient care.

The three authors of the Sydney Team wish to acknowledge the assistance of the other members of the team and including: Dr Christine Wallace (Head of the clinical department of Oral Restorative Sciences), Professor Ali Darendeliler (Head of Orthodontics, Faculty of Dentistry), Dr Evelyn Howe (Clinical Psychologist).

Although 10 teams were selected from established centers to work with the Sydney team, it is recognized that other teams could have been included. The groups were selected on the basis of personal knowledge by the Sydney team in the different locations, and selection was not meant to be exclusive. Since the completion of this part of the study, other team leaders have indicated their interest in the field, and additional teams would be welcome to join the proposed multicenter study, which is described in Part 2.

The authors reported no conflicts of interest related to this study.

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