Oral health and 22q11 deletion syndrome: thoughts and experiences from the parents' perspectives

GUNILLA KLINGBERG¹, ULRIKA HALLBERG² & SÓLVEIG ÓSKARSDÓTTIR³

¹Mun-H-Center, National Orofacial Resource Centre for Rare Disorders, and Department of Pediatric Dentistry, The Sahlgrenska Academy at the University of Gothenburg, Göteborg, Sweden, ²Nordic School of Public Health, Göteborg, Sweden, and ³Department of Paediatrics, The Sahlgrenska Academy at the University of Gothenburg, Queen Silvia Children's Hospital, Göteborg, Sweden

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Background. 22q11 deletion syndrome (22q11DS) is one of the most common multiple anomaly syndromes, and many dentists are likely to meet patients with the syndrome. Odontological research has focused on describing and analysing conditions/concepts based on the current state of knowledge within the dental profession. Yet, these research topics are not necessarily the most important issues for the patients.

Aims. To explore and describe, by use of Grounded theory, parents' experiences of oral health issues and needs for dental care in their children with 22q11DS.

Introduction

Although the 22q11.2 deletion syndrome (22q11DS) is one of the most common multiple anomaly syndromes with an incidence of about one per 4000 newborns^{1–3}, it is still under-diagnosed⁴. Many dentists are likely to meet patients with the diagnosis and it is thus important that foremost specialist in paediatric dentistry and special care dentists are familiar with 22q11DS.

The 22q11DS is caused by a hemizygous deletion of the q11.2 band of chromosome 22. In the beginning of the 1990s, this micro deletion was detected in the majority of patients with DiGeorge syndrome and velocardiofacial syndrome, and it became clear

Correspondence to:

Gunilla Klingberg, Folktandvården Västra Götaland, Mun-H-Center, National Orofacial Resource Centre for Rare Disorders, Odontologen, Box 7163, SE 402 33 Göteborg, Sweden. E-mail: gunilla.klingberg@vgregion.se **Design.** Twelve parents from different regions in Sweden were interviewed. Analyses were carried out according to Grounded theory.

Results. Parents recognised good oral health as important for the wellbeing of their children. Oral health was a concern and the parents described the fight for this as struggling in vain for good oral health in their child.

Conclusions. Parents not only described their children's oral health as important but also hard to gain. Thus, it is important that all patients with disabilities, regardless of whether there is a defined medical diagnosis or not, are identified and well taken care of in the dental care system.

that the original descriptions of these syndromes included various presentations of the same syndrome⁵. Nowadays the collective term 22q11 deletion syndrome (22q11DS) is commonly used for the syndrome although the original labels also are used.

The condition has an autosomal dominant inheritance, where approximately 80-90% of the deletions occur *de novo*⁵. The phenotypic spectrum is wide and includes characteristic features such as congenital heart defects, velopharyngeal insufficiency with or without cleft palate, infectious problems due to thymic hypoplasia, feeding difficulties, hypocalcaemia/hypoparathyroidism, developmental delay, learning disabilities, and behavioural abnormalities. Other malformations can occur, for example, urinary tract, skeletal, and neurological. The clinical appearance varies considerably and several patients show a complex medical picture^{2,6-9}. Yet, it is common to find that a patient has several less severe symptoms, but added up these symptoms cause

substantial disability¹⁰. The diagnosis is usually established at an early age for children with cardiac defects, whereas individuals without these problems have a risk of diagnostic delay. Developmental delay, learning difficulties or speech–language impairment are the most frequent symptoms leading to diagnosis in the latter group⁸.

Previous clinical and histological studies have reported that high frequency of dental aberrations, foremost enamel hypoplasia and hypomineralisation, as well as high caries prevalence in patients with 22q11DS^{11,12}. Apart from dietary and oral hygiene problems, impaired saliva secretion and altered composition regarding electrolytes and proteins are plausible explanations for the cariologic problems seen in many patients with 22q11DS¹³.

The clinical impression is that oral health issues are important for this group of patients, however, studies on this topic are lacking. Odontological research has focused primarily on describing and analysing oral tissues, functions, conditions, or concepts based on the current state of knowledge within the dental profession. Yet, these research topics are not necessarily the most important issues for the patients. This is especially true when meeting patients with more rare conditions. For example, a previous study on oral aspects of rare conditions showed that parents paid more attention to how dental care was provided than to the oral health issues¹⁴. The child's disability affected all aspects of family life and living, and oral health issues were not really on the agenda. The same pattern was revealed when interviewing patients with disabilities¹⁵. This is a problem as medical conditions/disabilities increase the risk for oral health problems. If the patients or their proxies do not recognise oral health issues as important, professionals have an even greater responsibility to provide good oral care for these groups of patients in order to ensure them oral health on equal terms with the rest of the population. As 22q11DS is a relatively common diagnosis where oral health problems are known, this study provides an opportunity to study this complexity.

The aim of this study was, therefore, to explore and describe, by use of Grounded

theory, parents' experiences of oral health issues and needs for dental care in their children with 22q11DS.

Method

Grounded theory

In this study the inductive qualitative research method Grounded theory (GT) was used¹⁶. GT is especially suitable when studying a phenomenon that is previously sparsely studied. It is also suitable when one wants to gain new knowledge on a previously studied area. The aim of a GT study is to deepen the knowledge and understanding of a phenomenon and to create a theory that can explain and/or predict this phenomenon. Grounded theory is rooted in symbolic interactionism and includes the assumption that meaning is constructed and changed by interactions among people¹⁷. GT can also be seen as a product of pragmatism and factor analysis¹⁸.

Three versions of GT have emerged; classical GT¹⁶ which ontologically is closer to positivism, because it is implicitly assumed that there is a real reality out there that can be discovered through emerging categories without being affected by the 'objective' researcher¹⁹. Modified GT, where Strauss and Corbin²⁰ further developed the method towards a postpositivistic standard and also presented a coding paradigm for facilitating the analysis process. The basic methodological principles are maintained, but Strauss and Corbin²⁰ state that the researcher interprets data rather than seeing categories emerging from the data. Later, Charmaz²¹ developed a constructivist Grounded theory aimed at giving an interpretative understanding of the studied area. Ontologically, Charmaz^{21,22} assumes that there are multiple realities, rather than the one and only 'real reality', and argues that 'grounded theories are constructions of reality'. In this study we have been influenced by the classical version of GT¹⁶.

Participants and procedure

Efforts were made to cover different aspects of the 22q11 DS in terms the child's age at

diagnose, age of the child at time of interview, living area, and family structure. Parents were chosen as informants as we wanted to explore viewpoints on oral health issues regardless of age. Parents interested in participating were recruited in collaboration with the Swedish patient organisation and the 22q11DS team at Sahlgrenska University Hospital in Göteborg. The first couple of participants were recruited this way. A selection of participants were then made to include parents whose children represented different ages, and also different ages at the time of disclosure of diagnosis. This was important as the possibilities to diagnose 22q11 DS as well as the awareness/knowledge about the diagnosis have developed over the passed 10-15 years. Further, participants were recruited from other parts of Sweden (cities, smaller towns, and country side living) in order to reflect possible differences in medical and dental care, both mothers and fathers were included, and the participants represented traditional families with two parents as well as single parent families. Twelve parents (2 fathers, 10 mothers) living in different regions of Sweden participated. None of the parents had 22q11DS. Their children diagnosed with 22q11DS (five girls/females, seven boys/males) were 2-40 years of age when the interviews were conducted. Their mean age was 18.5 years (two preschool age, two school age, two teenagers, and six adults). The children's age at disclosure of 22g11DS varied from first week of life to over 30 years of age. They had a mean age of 11.5 at diagnosis (five under the age of 2, four school age or teenage, three adults).

Open, tape-recorded interviews were carried out by one of the authors (U.H.) in a conversational style in the informant's homes or via telephone as requested by the informants, lasting up to 90 min. Telephone interviews are frequently used to collect data in qualitative research. Despite that the nonverbal language get lost, there is at least one major advantage; the possibility to interview participants at a long distance. Thereby, the study group gets more heterogeneous which also contributes to the transferability of the results²³. An interview guide was used and comprised the informants' experiences, prioritisation, thoughts and feelings on oral health care and treatments for their children. The informants had the opportunity to raise questions of relevance to them and the interviewer asked relevant follow up and probing questions during the interview. Data collection and analyses were conducted simultaneously and continued until new interviews did not provide any additional information, that is, so called saturation was reached.

Analysis

The data were analysed in a hierarchical coding process, that is, open and selective coding¹⁶. Open coding means line-by-line reading of the data and asking questions to the data such as: 'What is expressed here? What is the meaning of this?' By this, the substance of the data was captured and segmented into substantive codes, which were labelled concretely either with words from the informants, in vivo codes or by words from the authors, in vitro codes. At the end of the process of open coding, codes with similar contents were grouped together in categories. These categories were given more abstract labels than the codes belonging to it. In the selective coding a corecategory, central in the data, emerged describing what it was all about, and the categories were saturated with additional information, assessed by new interviews or added by re-coding previously assessed data, that is, theoretical sampling. During the entire process of analysis, ideas, preliminary assumptions, and theoretical reflections were written down in notes or 'memos'¹⁶. The results were presented to and discussed with a group of parents and individuals with 22q11DS as means for validation. The audience all recognised their own situation and struggled when hearing about the results. This is one way to validate research results, to go back to the participants and let them decide whether they find the results reasonable or not^{24} .

Results

During the analysis process a core category emerged labelled struggling in vain for good oral health in their child which illustrates the parents' concern and struggle for good oral health in their children. It was also obvious that receiving a diagnosis was important to gain legitimacy for the children's oral health problems in the dental office.

But...well, one becomes chastened with time. I've been doing this for 28 years. All I can say is that I've done absolutely everything in my power. And so says those around me too. I have spoken to God and to everyone I know, and ...

The children's oral health was of concern to the parents. Many parents described discoloured teeth and bad enamel as oral symptoms of their children's diagnosis, and described how this increased the risk for dental caries. In addition, carious lesions led to discoloured teeth which affected the children's smiles and appearances in a negative way. Many children were unable to understand the importance of taking good care of their teeth on a daily basis. This was a major problem especially in the older or grown up children; individuals that were really expected to take responsibility for their oral health. The parents were engaged on a full time basis (as parents, not as professionals) in the care of their child but there were often more acute physical and psychological problems that were regarded as more important than the oral health issues. Thus, the dental and oral health had to stand back for more general health problems. The knowledge and engagement from the dental health care professionals also varied from clinic to clinic or from dentist to dentist. Taken together this was seen as main barriers for gaining good oral health in their children. This is presented as a model in Fig. 1. These barriers are further described as categories below and selected quotations are indented to further illuminate the categories.

Heterogeneous caring competence

During the analysis, it became obvious that the informants had very different experiences regarding dental health care professionals'



Fig. 1. Model showing the core category struggling in vain for good oral health in their child in relation to the five categories. The struggle in vain can be described as a burden under the categories, and the categories constitute a barrier for gaining good oral health in the children with 22q11DS.

approach towards them and their child. Some of the informants described how the personnel were very understanding and seemed to have significant knowledge of the diagnosis and treatment needs. Some professionals also engaged more in the child's care than they were obliged to, such as transferring the child with them as a patient if they got a new job in another dental clinic. These parents felt secure and convinced that their children's oral health were in the best of hands. At the same time there were some informants that described how they had encountered dental health care professionals without any knowledge regarding the diagnosis, how it affected the oral health, or how to meet and treat their child. Some informants perceived the dentists blaming them as parents for the child's dental problems. This was more common among informants whose children had received the diagnosis at an older age. The disclosure of the diagnosis had sometimes led to a shift in attitudes from the dentists. Instead of seeing the child's oral health problems as a result of bad dietary and oral hygiene habits, they saw the child's dental problems as part of a medical diagnosis. Yet, still several parents had experienced how the dentists were unsure of how to treat the children and that they as parents had to take more responsibilities than they thought was fair. For example, some described how the choice of treatment strategies or modes of treatment were sometimes left for them to decide in situations where the dentists were unsure of how to deal with the child's dental problems or if the child did not cooperate with the treatment. Some informants also described how they sometimes had had no other choice than to hold the child by force in order for their child to have a dental examination or treatment. This was seen as violation both concerning the child and the parent. These children did not want to go to the dental health care clinic.

And when we get there it turns out that it is up to us, as parents, to decide if she is going to have a GA or if they should give her a tablet instead. And ... well, how are we supposed to know what's best? No, sorry but we cannot make this decision for you, they say... and okay... but what's best for my daughter?

Down prioritising oral health

The informants described how they had to prioritise medical issues or other psychological and physical problems in their children. Oral health was not the main priority. There always seemed to be general health problems that were more urgent or more imporfor their children's medical tant and psychological wellbeing. This included a wide range of problems from urgent heart surgery, immunological problems, to neuropsychiatric or psychiatric problems, to problems at school, or peer relations. The informants were aware of the problem of not being able to prioritise oral health issues, but they felt unable to do anything about it. They described having to take responsibility for scheduling appointments at the dental health clinics in order to make sure that the child should receive regular care and this was a responsibility they would like to hand over to the dental health care professionals instead.

Her teeth were not so important, in relation to her psychiatric and medical health. I have to say that ... it sort of ... it wasn't highly prioritised.

Keeping pace with daily oral care and acute oral problems

Some informants described their children having major dental caries problems despite careful daily oral health care owing to impaired enamel. Other informants lacked explanation from the dentists to why their children's teeth were discoloured. The informants described how they felt that they had nagged about the importance of brushing one's teeth carefully twice a day for the child's entire life. Many of the parents had continued to brush their children's teeth even until their children had reached their late teens. They had a strong wish to take extra good care of their children's teeth in order to avoid dental caries. In spite of this, their children developed new cavities over and over again. Some of the children had physical difficulties making it difficult to carry out oral hygiene procedures. Further, some of the children did not understand the importance of taking care of their teeth were not motivated, or lacked the energy to care about their oral health. The energy and time for oral health was focused on oral hygiene and taking care of dental problems as they occurred.

... it wasn't the main concern ... apart from brushing her teeth twice a day and that we saw the dentist and made sure that any cavities were taken care of.

Supporting normalisation in the child

The children diagnosed with 22q11DS were described as expressing a wish to be like everyone else also regarding oral health issues and oral health care. They did not want their teeth to be discoloured; instead they wanted to have good looking teeth like everybody else. The informants described how their children felt sad for their sometimes yellow or brownish looking teeth. The financial part was difficult for many of the grown up children. Many did not have regular jobs owing to problems related to the diagnosis, and the costs for dental care were not bearable. In addition, concerning oral self care the children wanted to be like others. Several parents described that they had bought powered toothbrushes in order to facilitate tooth brushing for the child as well as the parent, who often helped the child brushing the teeth. Many children reacted negatively to the powered toothbrush at first, and thought that they had been given this kind of toothbrush owing to their disability or diagnosis. Consequently, they wanted to keep their old toothbrush instead. After the parents had explained to them that many people use powered toothbrushes the children accepted this.

I bought one of those ... I mean he has had it for several years now... a powered toothbrush. He is averse to all those things... things that ... might make it easier for him because he thinks that... well, he thinks it has to do with his diagnosis. He kind of... just wants to be normal. But then I tell him that he's not the only one using a powered toothbrush, everyone does. Well almost. Then, he says like ... well he thinks it is OK to use it after all.

Having difficulties in setting limits

The informant described the dilemma of parenting a child with a disability. At the same time as the parents had to be supporting and encouraging, they also had to set up rules and boundaries. The informants frequently described problems related to feeding, diet, and eating habits. For example, some children wanted to eat a lot of candy or drink soft drinks every day. The parents knew that this was really negative for their children's dental health, especially as the children had an increased risk for dental caries. The same parents described how they had problems in being firm towards the child regarding restrictions. In many situations this need for sweets had been helpful for the child, for example, after complicated heart surgeries when it was important to make the child eat and drink something. In addition, many of the children had suffered from nutrition problems, difficulties to eat, etc., and in these situations it had been important to feed the child with something they liked and wanted to eat. Some informants described how their children had been addicted to a certain kind of lemonade and experienced head ache when the parents tried to be firm and restrictive towards the lemonade consumption. The informants also described how their children forgot not to eat or drink after tooth brushing at night.

There is one thing she really likes a lot, and that is drinking Coke. It has been both a blessing and a curse. And she is well aware of that. When she had her heart surgery, the very first thing she asked for, already at the ICU, was Coke and pizza.

Discussion

This study has presented a model showing that parents of children with 22q11DS recognise good oral health as important for the wellbeing of their children. Oral health was a concern and the parents described the fight for this as struggling in vain for good oral health in their child. They were well aware of the different oral health problems that are prevalent in 22q11DS and were especially worried about caries problems, discoloured teeth, expensive dental care for the grown up children, and for difficulties in motivating their children to good oral health habits including tooth brushing. A medical diagnosis was important to gain legitimacy for the oral health problems when seeing the dentist.

Having a child with a disability is known to imply stress on parents and family^{14,25,26}, and this study clearly showed that also oral health issues constituted a potential stressor. This is important knowledge for the dentists as it is essential that the dental staff understands and acknowledges that oral health problems in children with disabilities increase the burden on parents. Several oral health problems have been described in 22q11DS^{11–13}, and this information was accessible to the dental teams responsible for the dental care of the children in the study through, for example, PubMed. Still, several parents shared experiences of meeting of dental health care professionals who were not acquainted with the diagnosis at all. For some families, the parents had been blamed for the dental health problems in the child with 22q11DS. The dental teams had suggested improper diet or oral hygiene and did not acknowledge the problems that come with being a child with numerous infections, need for hospitalisation, etc., as concomitant factors for the dental problems. Often the more holistic understanding did not occur until the diagnosis was established and the dentist had received more information about the syndrome.

diagnosis implied legitimacy The and understanding for the oral health issues. This is an interesting and an important finding; obviously, the parents perceived that their children's oral health problems were approached by the dentists in a different way once the medical diagnosis was disclosed although the dental problems remained the same. There is really no good explanation for this. From a professional point of view, all patients should be cared for and treated equally based on their dental and oral health status. Looking at the medical history of these patients, all of them had medical problems such as, congenital heart defects, immunological problems, etc. and did not pass as healthy patients. The disclosure of 22q11DS could not possibly have changed that picture. From that perspective, it might even be alarming if patients with multiple but more isolated medical problems are not acknowledged as patients needing special attention and understanding from the dental team. These problems were captured in the category heterogeneous caring competence.

Yet, there could be plausible explanations for the different approaches from the dental teams. One concerns strategies when meeting patients with different medical conditions or syndromes. Previous research suggested that dental personnel have very different attitudes and that, for example, professional commitment, experience, training, knowledge, and organisation and work environment are important²⁷. These factors also reflect educational issues as there is still limited training and lecturing about special care patients in undergraduate education for dentists^{28,29}.

Another possible explanation concerns the lack of information on dental or oral health

aspects in medical conditions. For the majority of rare diagnoses, there are no studies at all on specific oral health issues related to the diagnosis. It is therefore plausible that many patients with rare conditions (and their families) frequently come across oral health professionals with no, or limited, knowledge about oral health aspects of the specific diagnosis. This is of course far from optimal. For the majority of disabilities the diagnosis as such, that is, the underlying cause of a condition, does not affect dental hard tissues, mucosa, or saliva, and there should not be an increased risk for dental health problems from the beginning. Yet, disabilities affect many other aspects of life that in turn may lead to oral health problems, and this must be recognised by professionals while meeting these patients in the dental office. Many conditions go hand in hand with learning disabilities and/or neuropsychiatric problems. Although these problems do probably not increase the risk for dental health problems in childhood per se, they often impose cooperation and behaviour problems that make any dental treatment a challenge for the dentist. Thus, in a more long term perspective, it is likely to find an impaired dental health in these groups of patients³⁰. Other aspects likely to affect oral health are dietary habits, medication, feeding problems, hypotonic muscles, etc. Based on this, just knowing a (child) patient has a disability or a medical condition should ring a bell to the dental team that oral health could be at risk.

Although there might be limited literature available for many diagnoses, and even sparser when it comes to oral health issues, all dental teams need to work out strategies to ensure quality in the care for patients with disabilities. These could preferably include seeking medical and dental information about the diagnosis, allowing more time for anamnesis, and communication with patients (and parents) to learn to know the individual patient better, ensuring sufficient time for the appointments in order to reduce the risk of stress during treatment, and continuous education in dental care for patients with disabilities for all members of the dental team.

Several parents in this study perceived that their children had more dental health

problems than others, and that their children not always received comprehensive dental treatment. Only limited research has been conducted in this area. The findings from this study are supported by reports on parents of children with Down syndrome in the UK³¹. Parents in that study expressed that their children had unmet dental treatment needs, and that they wanted more preventive dental measures and collaboration with other health professionals. The authors also concluded that the parents urged for sympathetic dentists who were good communicators and well informed about their child's diagnosis³¹, findings that support the results in this study.

In this study, the children, whose parents were interviewed, had a wide age range. Some had received their diagnosis at an early age; others were older when 22q11DS was disclosed. The age differences implied a variation in experiences and these were reflected in the interviews. The different ages of the children were essential in order to gain a more complete understanding of parents' perceptions of the oral health issues. It was thus important to learn also from parents of older children as their experiences are useful for understanding how meeting and caring for the child patients of today can be improved.

There is always a risk for bias also in a study using qualitative methods in terms of the information being more easily gathered from people that have an interest or knowledge about the subject. For this study, the strategic sampling tried to embrace informants who were not interested in oral health aspects of the syndrome. Therefore, the two categories pinpointing the fact that oral health issues have to stand back for medical problems and managing the everyday life in a family where one's daughter or son happened to have a disability (Down prioritising oral health and Keeping pace with daily oral care and acute oral problems) are of special interest. The results are supported by previous reports of oral health issues being marginalised in the wider context of health and daily life for people with disabilities^{14,15}. These findings are essential reminders for dental health care professionals that dental issues are not top priority for the patients/families.

This does not mean that oral health is not important, but the life situations for these families imply that they have to focus on other aspects of health and wellbeing. As disability may increase the risk for oral health problems, the fact that the individual patient and her/his family have other items on their main agenda implies higher demands on the dental team in order to prevent these problems.

Parenting a child with a disability puts strain on the family, and differs from parenting a child without disability $^{32-34}$. Regardless of the age of their child, informants in this study described the dilemma of supporting the child almost endlessly and at the same time having to work hard to be consistent regarding boundaries. This was captured in the section 'Having difficulties in setting limits'. Much of this concerned eating habits, food, and nutrition which are closely related to dental health. Parents described a wish to have an authoritative style in their parenting, but that this was difficult, a finding in line with previous studies³⁴. They really wanted the best for their child, and were well aware of the potential dental problems related to poor eating habits. But at the same time they had to consider other health issues in case of children with nutritional or eating problems. Sometimes allowing sweetened beverages or fast food was a way to avoid conflicts, and on some occasions they just felt a need to spoil their son/daughter for a while. All these aspects are understandable and should not be looked upon as part of a permissive parenting style where the parents do not care about diet or dental health. Instead, it may serve as a good example of the complexity and challenges when parenting a child with disability. From a dental health point of view the parenting aspects may be problematic. But just informing about good or ideal dietary or oral hygiene habits will not improve the oral health for the person with 22g11DS. Instead, the dental treatment plan has to take parenting dilemmas into consideration and modify communication and preventive strategies to the individual patient in order to prevent oral health problems. It is advocated that treatment include more chair side prevention than what is usually needed for other healthy patients with similar oral health conditions. As many patients with 22q11DS have problems cooperating with dental treatment owing to cognitive impairment, neuropsychiatric problems, etc., prevention is even more important in order to maintain good oral health.

The descriptions of how the children expressed that they wanted to be like everyone else (found in the section 'Supporting normalisation in the child') also regarding oral heath was reassuring to find. It implies that oral health is important to people with 22g11DS. The sons and daughters were aware of problems with decay and discoloration, and this bothered them. At the same time, they had difficulties taking full responsibility for their dental health, for example, many of the informants described their children as having problems managing their oral hygiene. The fact that several of them had increased costs for their dental care compared with others of the same age was pointed out as especially unfair and problematic. The parents expressed that the dental problems were not caused by the son/daughter or family, but rather an effect of the syndrome as such. Therefore, they demanded extended help from the dental teams and society (in terms of financial support) in order to ensure that their child, regardless of age, should have the same possibility as others to gain and preserve good oral health.

In conclusion, there are several risk factors for impaired oral health in individuals diagnosed with 22q11DS. Parents not only described their children's oral health as important, but also hard to gain, as something they had to fight for against all odds. This fight included achieving proper dental care from dental teams with knowledge about 22q11DS as well as the challenges that come with being a family with a child with disability. Oral health had to compete with many other issues that were on the agenda. Problems of gaining legitimacy for oral health issues, especially before the medical diagnosis was known, may increase the risk for impaired oral health in individuals with 22q11DS. Thus, it is important that all patients with disabilities, regardless of whether there is a defined medical diagnosis or not, are identified and well taken care of in the dental care system.

What this paper adds

- Insight into how parents of children with disabilities perceive that they have to fight for good oral health in their children.
- Knowledge about the oral health dilemmas in patients with 22q11 deletion syndrome.
- Parents' descriptions of how a defined medical diagnosis is important to gain legitimacy for oral health problems at the dental office.

Why this paper is important to paediatric dentists

- Learning from parents' perceptions of the children's dental care can help paediatric dentists to further develop and improve dental care for children with disabilities.
- It is important to recognise oral health problems in children with disabilities regardless of the underlying diagnosis.

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References

- 1 Devriendt K, Fryns JP, Mortier G, van Thienen MN, Keymolen K. The annual incidence of DiGeorge/ velocardiofacial syndrome. *J Med Genet* 1998; **35**: 789–790.
- 2 Botto LD, May K, Fernhoff PM *et al.* A populationbased study of the 22q11.2 deletion: phenotype, incidence, and contribution to major birth defects in the population. *Pediatrics* 2003; **112**: 101–107.
- 3 Óskarsdóttir S, Vujic M, Fasth A. Incidence and prevalence of the 22q11 deletion syndrome: a population-based study in Western Sweden. *Arch Dis Child* 2004; **89**: 148–151.
- 4 Shprintzen RJ. Velo-cardio-facial syndrome: 30 Years of study. *Dev Disabil Res Rev* 2008; **14**: 3–10.
- 5 Swillen A, Vogels A, Devriendt K, Fryns JP. Chromosome 22q11 deletion syndrome: update and review of the clinical features, cognitive-behavioral spectrum, and psychiatric complications. *Am J Med Genet* 2000; **97**: 128–135.

- 6 Yamagishi H, Srivastava D. Unraveling the genetic and developmental mysteries of 22q11 deletion syndrome. *Trends Mol Med* 2003; **9**: 383–389.
- 7 Ryan AK, Goodship JA, Wilson DI *et al.* Spectrum of clinical features associated with interstitial chromosome 22q11 deletions: a European collaborative study. *J Med Genet* 1997; **34**: 798–804.
- 8 Óskarsdóttir S, Persson C, Eriksson BO, Fasth A. Presenting phenotype in 100 children with the 22q11 deletion syndrome. *Eur J Pediatr* 2005; **164**: 146–153.
- 9 Hopkin RJ, Schorry EK, Bofinger M, Saal HM. Increased need for medical interventions in infants with velocardiofacial (deletion 22q11) syndrome. *J Pediatr* 2000; **137**: 247–249.
- 10 Óskarsdóttir S, Belfrage M, Sandstedt E, Viggedal G, Uvebrant P. Disabilities and cognition in children and adolescents with 22q11 deletion syndrome. *Dev Med Child Neurol* 2005; **47**: 177–184.
- 11 Klingberg G, Óskarsdóttir S, Johannesson-Lövsund E, Norén JG. Oral manifestations in 22q11 deletion syndrome. *Int J Paediatr Dent* 2002; **12**: 14–23.
- 12 Klingberg G, Dietz W, Óskarsdóttir S, Odelius H, Gelander L, Norén JG. Morphological appearance and chemical composition of enamel in primary teeth from patients with 22q11 deletion syndrome. *Eur J Oral Sci* 2005; **113**: 303–311.
- 13 Klingberg G, Lingström P, Óskarsdóttir S, Friman V, Bohman E, Carlén C. Caries-saliva properties in individuals with 22q11 deletion syndrome. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2007; 103: 497–504.
- 14 Trulsson U, Klingberg G. Living with a child with a severe orofacial handicap: experiences from the perspectives of parents. *Eur J Oral Sci* 2003; **111**: 19–25.
- 15 Hallberg U, Klingberg G. Giving low priority to oral health care. Voices from people with disabilities in a grounded theory study. *Acta Odontol Scand* 2007; **65**: 265–270.
- 16 Glaser BG, Strauss AL. *The Discovery of GROUNDED theory. Strategies for Qualitative Research.* Chicago: Aldine, 1967.
- 17 Mead GH. *On social psychology*. Chicago: University of Chicago Press, 1969.
- 18 Schreiber RS. The 'How to' of grounded theory: avoiding the pitfalls. In: Schreiber RS, Stern PH. (eds). *Using Grounded Theory in Nursing*. New York: Springer Publishing, 2001: 55–83.
- 19 Hallberg LR-M. The 'core category' of grounded theory: making constant comparisons. *Int J Qual Stud Health Well-being* 2006; **1**: 141–148.
- 20 Strauss AL, Corbin JM. Basics of Qualitative Research. Techniques and Procedures for Developing Grounded Theory. Thousand Oaks, CA: Sage, 1998.

- 21 Charmaz K. Grounded theory. In: Smith JA, Harre R, van Langenhove L. (eds). *Rethinking Methods in Psychology*, 2nd edn. London: Sage, 1995: 27–49.
- 22 Charmaz K. Grounded Theory. Objectivist and constructivist methods. In: Denzin NK, Lincoln YS. (eds). *Handbook of Qualitative Research*, 2nd edn. Thousands Oaks, CA: Sage, 2000: 509–535.
- 23 Kvale S, Brinkmann S. *Den kvalitativa forskningsintervjun*, 2nd edn (*The qualitative research interview*). Lund: Studentlitteratur, 2009.
- 24 Larsson S. Om kvalitet i kvalitativa studier (On quality in qualitative research). *Nordisk Pedagogik* (*J. Nordic Educ. Res.*) 1993; **13**: 194–211.
- 25 Seltzer MM, Greenberg JS, Floyd FJ, Pettee Y, Hong J. Life course impacts of parenting a child with a disability. *Am J Ment Retard* 2001; **106**: 265–266.
- 26 Seltzer MM, Almeida DM, Greenberg JS *et al.* Psychosocial and biological markers of daily lives of midlife parents of children with disabilities. *J Health Soc Behav* 2009; **50**: 1–15.
- 27 Hallberg U, Strandmark S, Klingberg G. Dental health professionals' treatment of children with disabilities: a qualitative study. *Acta Odontol Scand* 2004; **62**: 319–327.
- 28 Fenton SJ, Hood H, Fenton SJ *et al.* The American Academy of Developmental Medicine and Dentistry: eliminating health disparities for individuals with mental retardation and other developmental disabilities. *J Dent Educ* 2003; **67**: 1337–1344.
- 29 Casamassimo PS, Seale NS, Ruehs K. General dentists' perceptions of educational and treatment issues affecting access to care for children with special health care needs. *J Dent Educ* 2004; **68**: 23–28.
- 30 Gabre P, Martinsson T, Gahnberg L. Longitudinal study of dental caries, tooth mortality and interproximal bone loss in adults with intellectual disability. *Eur J Oral Sci* 2001; **109**: 20–26.
- 31 Kaye PL, Fiske J, Bower EJ, Newton JT, Fenlon M. Views and experiences of parents and siblings of adults with Down Syndrome regarding oral healthcare: a qualitative and quantitative study. *Br Dent J* 2005; **198**: 571–578.
- 32 Macias MM, Saylor CF, Rowe BP, Bell NL. Agerelated parenting stress differences in mothers of children with spina bifida. *Psychol Rep* 2003; **93**: 1223–1232.
- 33 Deatrick JA, Knafl KA, Walsh M. The process of parenting a child with a disability: normalization through accommodations. *J Adv Nurs* 1988; **13**: 15–21.
- 34 Woolfson L, Grant E. Authoritative parenting and parental stress in parents of pre-school and older children with developmental disabilities. *Child Care Health Dev* 2006; **32**: 177–184.

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