

ABSTRACT

The purpose of this study was to examine oral health needs and dental care in individuals with trisomy 18 and trisomy 13 (full, mosaic, partial and other, mixed types). Primary feeding method was also examined. Data was collected from a parent-completed, mixed method survey (TRIS Survey). Mean age in months was 120.2 (range 38 to 394 months) and 133 (range 36 to 405 months), respectively, for trisomy 18 and trisomy 13 individuals. Results indicated the majority of individuals received routine dental care from their family dentist. Approximately 80% in both groups needed some form of specialized dental care. Close to 25% and 30% of trisomy 18 and trisomy 13 individuals, respectively, required hospital admission for specialized dental care. Responses indicated the presence of excessive plaque and tooth decay across the groups with a higher incidence for individuals with trisomy 13. Although not the primary form of intake, over half of the individuals received oral feedings. Implications for dental care and management are provided along with the need for additional research to confirm or disconfirm this study's findings.

KEY WORDS: rare syndromes, trisomy 18, trisomy 13, feeding method, dental care, oral health needs

Oral health needs in individuals with trisomy 18 and trisomy 13: Implications for dental professionals

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Introduction

The literature offers limited information concerning oral health needs for individuals with rare trisomy conditions such as trisomy 18 and trisomy 13. Specifically, available articles on the topic present case studies or small samples with results that cannot be generalized while most large scale studies focus on survival, not treatment and with descriptions of cardiac defects and associated medical concerns but not oral health needs.¹⁻⁴

With recent reports of increased longevity,⁵⁻⁷ it is imperative to delve further into this area in order to identify common oral health needs and recommendations for dental care. What is needed is a more in-depth examination of the unique oral health needs and dental care necessary for optimal outcomes in these groups. The purpose of this study is to provide data from larger samples to informal dental professionals.

Prevalence rates are approximately one in 6,000 to 8,000 live newborns with trisomy 18 and one in 7,000 to 10,000 for trisomy 13. Respectively, they are the second and third most common autosomal conditions. Notably, individuals with trisomy 18 and trisomy 13 often experience cardiac and respiratory difficulties such as presence of ventricular septal defect (VSD) and need for supplemental oxygen or placement of a tracheotomy due to a small or malformed airway, respectively.^{8,9} There is also a confluence with reflux and a high

incidence of tube feeding.^{8,10} Cleft lip and/or palate have also been identified in these groups along with evidence of micrognathia and small mouth size.^{10,11} Individually and collectively, these medical conditions and physical characteristics can impact oral health needs and provision of basic and specialized dental care.

Available case reports describing oral health needs of individuals with trisomy 18 emphasize treatment related to excessive plaque and gingival inflammation and recession.¹²⁻¹⁵ Information is also offered about a 16 years old with trisomy 13 presenting with tooth decay, enamel erosion and bruxism.¹⁶ Treatment was provided under general anesthesia. Also, antibiotics and/or local anesthesia have also been noted as sometimes necessary prior to treatment. It is important to note mixed outcomes with anesthesia use in individuals with trisomy 18 and trisomy 13.^{9,17,18} Overall, the authors recommend proactive preventative care.

The 16 years old with trisomy 13 was also noted with oral feeding as the primary means of intake rather than enteral (tube) feedings.¹⁶ Most information on individuals with trisomy 18 or trisomy 13 only note feeding difficulties and/or identifies oral defects rather than treatment. For example, a sample with trisomy 13 was noted with a high incidence of cleft lip and palate ($n = 28$, 71%).¹⁹ Importantly, median survival was nine days with only one case living more than a year. As such, no dental management data is reported.

Taken together, these descriptions point to a need for more in-depth investigations toward identifying key oral health concerns and treatment for individuals with the trisomy 18 and trisomy 13 phenotypic and genotypic profiles. The resulting data can assist dental professionals to develop care and treatment plans to address common needs. To the authors' knowledge, this article is the first to offer information to inform short and long term oral health needs and dental care for individuals with trisomy 18 and trisomy 13. Feeding method data is also included as available research only highlights feeding difficulties.

Methods

The Tracking Rare Incidence Syndrome (TRIS) project began in 2007. Data is collected via parent-completed surveys focusing on the prenatal, neonatal and perinatal periods as well as past and current health and medical needs and corresponding interventions (e.g., surgery). Dissemination of results to interested audiences via articles and presentations has been ongoing since the project's inception.

Instrumentation

The TRIS Survey is completed by parents with a child 2 months of age or older. Child does not have to be living at time of survey completion. The Modified TRIS Survey was developed for parents with children living 60 days or less or still-born. The Modified TRIS Survey includes a subset of TRIS Survey items.

Survey access is via an online portal accessible through the TRIS project's webpage. Paper copies are provided by request and returned by postal mail in a stamped, self-addressed envelope. Data is then manually entered into the project's database.

The TRIS Survey is composed of three parts. Part I includes items on labor and delivery, suspected and presenting medical conditions, postbirth course and hospital discharge. There are also seven demographic items. Part II examines family support (56 items). Part III collects data related to a range of health needs and medical interventions including surgeries and medications (61 items).

Feeding method, oral health and dental care items are included in Part III. Each of the five items has subitems. For example, the one of the items asks, "What type(s) of dental problems has your child experienced?" Participants then review a list including too few teeth and excessive plaque and respond with a "Yes" or "No" to each option. The item asking for types of oral intake and enteral (tube) feeding is similarly structured. Remaining items ask for use of sedation and antibiotics during routine and specialized dental care.

Procedure

Approval was received from the Human Subjects Committee at Southern Illinois University Carbondale prior to the start of data collection.

Since the start of the project, participants were and continue to be have been recruited through announcements to rare trisomy-related websites, listservs and via articles in the Support Organization for Trisomy 18, 13 and related disorders (SOFT) newsletter. In addition, a Facebook page was launched in 2011 to attract participants and provide project updates. In 2013, a series of blog entries about the project were requested by the Global Genes project (see <http://globalgenes.org/raredaily/giving-a-face-to-trisomy-18-debbie-elaborates-on-this-rare-condition/> for an example). Recommendations from participants are another avenue for recruitment

(e.g., Facebook groups, attendance at annual SOFT Conference).

Participants complete a pre-enrollment form online with name, phone number, state/province and country, email address and child's name, date of birth (and date of death, if appropriate) and trisomy type. Within approximately 48 hours, the TRIS project Research Coordinator emails a login and password to access either the TRIS Survey or TRIS Modified Survey instrument to the email address provided on the pre-enrollment form. The log in and password is used for the duration of project participation.

TRIS project staff is able to view participant progress since the survey can be accessed multiple times on the TRIS project website prior to completion (participants are minded to save additional data prior to leaving the survey). An initial email reminder is sent when a survey is not completed within 1 month of enrollment. Additional reminders are sent every 2 to 3 months. At survey completion, TRIS project staff receives a completion notice by email. The server housing the TRIS project surveys and data is updated and archived daily.

Participants

The TRIS project database was examined for individuals meeting the following criteria: diagnosis of a trisomy 18 or trisomy 13 type, minimum age of 36 months at TRIS Survey completion and living at time of completion. The latter requirement stems from the purported later eruption of teeth in individuals with genetic syndromes (see de Queiroz *et al.*, 2007) compared with genetically typical children who have *all* primary teeth by approximately 30 to 33 months of age (<http://www.mouthhealthy.org/en/az-topics/e/eruption-charts.>)

It is of import to note that individuals with trisomy 18 and trisomy 13 often experience cardiac and respiratory conditions requiring immediate and/or ongoing attention during the first 3 years of life. Feeding issues are also often present. Coupled with late eruption, these medical factors also contribute to a limited focus on dental care and treatment unless a specific need arises.

Table 1. Demographic data at time of TRIS Survey completion.

Demographic variables	Trisomy 18 types (n = 51)	Trisomy 13 types (n = 44)
	Mean (±SD) Range	Mean (±SD) Range
Child's age	120.4 months (±88) Range: 38-394 months	133 months (±92.1) Range: 36-405 months
Mother's age at birth	32.8 years ^a (±6.2) Range: 22-44 years	31.1 years ^b (±6) Range: 18-43 years
Father's age at birth	35.1 years ^c (±6.4) Range: 23-48 years	33 years ^d (±6.8) Range: 23-52 years
	n (%)	n (%)
Marital status		
Single	2 (3.9%)	1 (2.3%)
Long-term relationship	1 (2.0%)	1 (2.3%)
Partnered	0 (0%)	1 (2.3%)
Married	40 (78.4%)	36 (81.8%)
Separated	3 (5.9%)	1 (2.3%)
Divorced	2 (3.9%)	4 (9.1%)
Widowed	1 (2.0%)	0 (0%)
Education level		
<6 years	1 (2.0%)	0 (0%)
7-9 years	0 (0%)	3 (6.8%)
10-12 years	6 (11.8%)	12 (27.3%)
13-16 years	17 (33.3%)	16 (36.4%)
17-20 years	21 (41.2%)	8 (18.2%)
>20 years	4 (7.8%)	5 (11.4%)
Income level^e		
Low	3 (5.9%)	7 (15.9%) ^g
Medium	^f 40 (78.4%)	32 (72.7%)
High	6 (11.8%)	4 (9.1%)

^an = 49.
^bn = 43.
^cn = 48 (three cases with trisomy 18 were adopted).
^dn = 43 (one case with trisomy 13 was adopted).
^eIncome data is collected by level not dollar amounts due to the international scope of the project; most participants lived in the U.S. at the time of survey completion but eight cases with trisomy 18 (two in Australia, three in Canada, one in England, one in Norway, and one in Scotland) and four with trisomy 13 did not (two in Australia, one in Canada and one in Portugal).
^fn = 49.
^gn = 43.

As of April 15, 2015, 351 TRIS Surveys were completed. Of the total, 95 individuals meeting the above criteria were identified (27.1%). Table 1 provides demographic results of the individuals and participants (parents completing the TRIS Survey). All individuals were living at the time of survey completion. Mean age in months was 120.2 (range 38 to 394 months) and

133 (range 36 to 405 months), respectively, for trisomy 18 and trisomy 13 individuals.

Most participants provided information for mother and father age at case's birth. Mean age for mothers was 32.8 years (trisomy 18) and 31.1 years (trisomy 13). Fathers were 35.1 and 33 years, respectively. Most participants identified as married (n = 40, 78.4% for

trisomy 18; n = 36, 81.8%). The majority in both groups completed at least 12 years of education and reported medium income level.

Data analysis

Survey item and demographic data are linked to each participant's unique TRIS project identification number. All data are downloaded to a spreadsheet in Microsoft Excel for visual inspection. The data is then copied into a database in SPSS.

Due to the nature of the items (binary Yes/No options), frequencies and percentages were computed. Nonparametric, descriptive statistical analyses were used with TRIS Survey demographic data. Frequencies, percentages as well as means and standard deviations were computed.

Results

The following sections provide results for feeding method, dental care and oral health needs. Each section provides the number of individuals with trisomy 18 and trisomy 13 described by the data.

Data represents 51 individuals with a type of trisomy 18 (14.5%) (Full = 32, Mosaic = 11, Partial = 2, Other = 6) and 44 with a type of trisomy 13 (12.5%) (Full = 18, Mosaic = 11, Partial = 5, Other = 10). "Other" refers to diagnoses with a combination of types such as partial trisomy 13p.

Feeding method

Results indicated that, at the time of survey completion, individuals with full trisomy 18 (n = 32) and full trisomy 13 (n = 18) were almost evenly divided between oral feeding and tube feeding (n = 15, 46.9%, n = 17, 53.1%; n = 8, 44.4%, n = 10, 55.6%, respectively). The percentage among mosaic, partial and other types varied but most in each category identified the use of oral feeding. Gastrostomy tube feeding was most prevalent across the groups. Only one child with full trisomy 13 required an intravenous port (central line) for feeding (refer to Table 2).

Table 2. Feeding methods at time of TRIS Survey completion.

	Oral feeding ^a	Tube feeding	Type of tube feeding ^b
	n (%)	n (%)	n (%)
Trisomy 18 types (n = 51)			
Full (n = 32)	15 (46.9%)	17 (53.1%)	16 (94.1%) g-tube 1 (5.9%) g-j tube
Mosaic (n = 11)	10 (90.9%)	1 (9.1%)	1 (100%) g-tube
Partial (n = 2)	2 (100%)	0 (0%)	n/a
Other (n = 6)	4 (66.7%)	2 (33.3%)	1 (50%) g-tube 1 (50%) g-j tube
Trisomy 13 types (n = 44)			
Full (n = 18)	8 (44.4%)	10 (55.6%)	7 (70%) g-tube 2 (20%) g-j tube 1 (10%) IV port
Mosaic (n = 11)	10 (90.9%)	1 (9.1%)	1 (100%) g-tube
Partial (n = 5)	4 (80%)	1 (20%)	1 (100%) g-tube
Other (n = 10)	8 (80%)	2 (20%)	2 (100%) g-tube
^a Oral feeding was not typically the primary intake method; included table food in various forms (pureed, chopped, etc.) and liquids from various cups (sippy, cut out, etc.).			
^b g-tube = gastrostomy tube; g-j tube = gastrostomy-jejunostomy tube; IV port = intravenous port/central line.			

Dental care

The following data describe responses to items asking about routine and specialized dental care for individuals with trisomy 18 and trisomy 13.

Routine dental care

Based on data from 93 individuals (trisomy 18, n = 50; trisomy 13, n = 43), the majority received routine dental care from their family dentist (t18 n = 45, 90%; t13

n = 34, 79.1%). In addition, approximately 75% in each group also saw a pediatric dentist. Sedation was identified for 22% in the trisomy 18 group (n = 11) and 20.9% in the trisomy 13 group (n = 9). Items also requested information about antibiotic use before and after routine dental care. Results indicated 30% of individuals with trisomy 18 (n = 15) and 11.6% (n = 5) with trisomy 13 needed antibiotics prior and less than 10% in

both groups after routine dental care (n = 4 and n = 1, respectively).

Specialized dental care

Data was available for 37 individuals with trisomy 18 and 34 with trisomy 13. Approximately 80% in both groups required specialized dental care (trisomy 18, n = 28; trisomy 13, n = 28). Sedation was required for 29.7% (n = 11) with trisomy 18 and 50% (n = 17) with trisomy 13. The need for hospital admission for specialized dental care was also requested. A total of 21.6% (n = 8) in the trisomy 18 groups and 29.4% (n = 10) with trisomy 13 required this option.

Results by specific trisomy type for routine and specialized dental care are provided in Tables 3 and 4.

Oral health needs

Available data on specific oral health needs indicated the presence of excessive plaque and tooth decay across the groups (trisomy 18, n = 34; trisomy 13, n = 33). For the sample with trisomy 18, 35.5% (n = 12) were identified with excessive plaque and 26.5% (n = 9) with tooth decay. Results were slightly higher for individuals with trisomy 13. Specifically, 42.4% (n = 14) with excessive plaque and 39.4% (n = 13) with tooth decay.

In addition, periodontal concerns were noted. For individuals diagnosed with trisomy 18, a total of five (14.7%)

Table 3. Routine dental care data (n = 93).

	Seen by family dentist	Seen by pediatric dentist	Sedated for routine dental care	Requires antibiotics prior to routine dental care	Requires antibiotics after routine dental care
	n (%)	n (%)	n (%)	n (%)	n (%)
Trisomy 18 types (n = 50^a)					
Full (n = 31)	29 (94%)	23 (74%)	6 (19%)	12 (39%)	4 (13%)
Mosaic (n = 11)	9 (82%)	7 (64%)	3 (27%)	2 (18%)	0 (0%)
Partial (n = 2)	1 (50%)	1 (50%)	0 (0%)	0 (0%)	0 (0%)
Other (n = 6)	6 (100%)	4 (67%)	2 (33%)	1 (17%)	0 (0%)
Trisomy 13 types (n = 43^b)					
Full (n = 18)	14 (78%)	14 (78%)	4 (22%)	2 (11%)	1 (6%)
Mosaic (n = 11)	8 (73%)	7 (64%)	1 (9%)	1 (9%)	0 (0%)
Partial (n = 5)	4 (80%)	3 (60%)	3 (60%)	0 (0%)	0 (0%)
Other (n = 9)	8 (89%)	7 (78%)	1 (11%)	2 (18%)	0 (0%)
^a No data provided for one case with full trisomy 18.					
^b No data provided for one case with trisomy 13 "other."					

Table 4. Specialized dental care data (n = 71).

	Required specialized dental care	Sedated for procedures	Requires antibiotics before procedures	Requires antibiotics after procedures	Requires hospital admission
	n (%)	n (%)	n (%)	n (%)	n (%)
Trisomy 18 types (n = 37^a)					
Full (n = 25)	19 (76%)	6 (24%)	7 (28%)	3 (12%)	5 (20%)
Mosaic (n = 7)	5 (71%)	3 (43%)	1 (14%)	0 (0%)	1 (14%)
Other (n = 5)	4 (80%)	2 (40%)	0 (0%)	0 (0%)	2 (40%)
Trisomy 13 types (n = 34^b)					
Full (n = 14)	11 (79%)	7 (50%)	2 (14%)	2 (14%)	3 (22%)
Mosaic (n = 8)	7 (88%)	3 (36%)	1 (13%)	0 (0%)	2 (25%)
Partial (n = 4)	3 (75%)	3 (75%)	0 (0%)	0 (0%)	2 (50%)
Other (n = 8)	7 (86%)	4 (50%)	1 (13%)	1 (13%)	3 (38%)

^aNo data provided for seven cases with full trisomy 18, four cases with mosaic trisomy 18, two cases with partial trisomy 18, and one with trisomy 18 "other."
^bNo data provided for four cases with full trisomy 13, three cases with mosaic trisomy 13, one case with partial trisomy 13, and two cases with trisomy 13 "other."

Table 5. Oral health issues (n = 67).

	Too few teeth	Too many teeth	Excessive plaque	Tooth decay	Periodical problem/gum sensitivity or disease
	n (%)	n (%)	n (%)	n (%)	n (%)
Trisomy 18 types (n = 34^a)					
Full (n = 21)	2 (10%)	3 (14%)	7 (33%)	5 (24%)	2 (10%)
Mosaic (n = 7)	1 (14%)	1 (14%)	3 (43%)	2 (29%)	2 (29%)
Partial (n = 1)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Other (n = 5)	0 (0%)	0 (0%)	2 (40%)	2 (40%)	1 (20%)
Trisomy 13 types (n = 33^b)					
Full (n = 14)	0 (0%)	0 (0%)	6 (43%)	4 (29%)	0 (0%)
Mosaic (n = 6)	1 (17%)	2 (33%)	3 (50%)	2 (33%)	0 (0%)
Partial (n = 4)	0 (0%)	0 (0%)	2 (50%)	3 (75%)	3 (75%)
Other (n = 9)	1 (11%)	0 (0%)	3 (33%)	4 (44%)	1 (11%)

^aNo data provided for 11 cases with full trisomy 18, 4 cases with mosaic trisomy 18, 1 case with partial trisomy 18, and 1 with trisomy 18 "other."
^bNo data provided for five cases with mosaic trisomy 13, one case with partial trisomy 13, and one case with trisomy 13 "other."

exhibited this oral health condition and four with trisomy 13 (12.1%). Results by specific trisomy type for oral health needs can be found in Table 5.

Discussion

Results identified several trends. Approximately half of the individuals with full trisomy 18 and full trisomy 13 received some nutrition via oral feeding. In addition, gastrostomy tube feeding was prevalent across both groups.

For routine dental care, data indicated that most individuals received

routine dental care from their family dentist. Most individuals also saw a pediatric dentist. Some individuals required antibiotics prior to routine dental care. Many required some form of specialized dental care including hospital admission for procedures. Finally, responses indicated the presence of excessive plaque and tooth decay across the groups with a higher incidence for individuals with trisomy 13.

These findings provide additional information not available in the literature. As stated earlier, feeding difficulties are often noted with little description of

feeding methods. Providing an overview of routine and specialized dental care for individuals with trisomy 18 and trisomy 13 can also assist dental professionals with general parameters to guide treatment plans (e.g., frequency of excessive plaque, need for antibiotics). Taken together, the clinical relevance of the results provided here is toward raising awareness of the oral health needs of these groups.

Confirmation of literature

One case report described the use of antibiotics and local anesthesia prior to

treatment provided in a dental office for an individual with trisomy 18¹⁵ and another with trisomy 13 receiving general anesthesia prior to treatment.¹⁶ Results with the samples described here included the use of antibiotics and anesthesia. The case with trisomy 13 was also noted to receive oral feedings. It is important to note that feeding method across the 95 individuals was part of this study. While not analyzed for co-occurrence with specific oral health needs (e.g., tooth decay), primary feeding method data offers contextual background for dental care.

Several authors note excessive plaque and/or periodontal concerns for individuals with these conditions.^{12–15} Specifically, enamel erosion is cited in an individual with trisomy 13.¹⁶ Again, similar findings were reported here for some individuals with trisomy 18 and trisomy 13.

Limitations

A number of limitations affected the results. First, the TRIS Survey only included five items asking for information about oral health and dental care. Survey items were intended to gather general information, not to be comprehensive as the first author's background is not in dentistry. For example, no items ask about presence of self-injurious behavior that may impact oral health and dental needs (e.g., fingers in mouth for extended periods of time, picking at gums). The goal of the study was an initial exploration of the topic.

It is also possible participants did not provide accurate data. This could be due to unfamiliar terminology and/or limited memory if there were no recent oral care or dental health issues. There was also a potential issue with "No" and blank responses to items. The authors reviewed available data multiple times to develop guidelines to address items without data (blanks). While desirable, it was not feasible to contact participants since some completed the survey more than 5 years ago and contact information had not been updated to locate them. Data described here should be interpreted with caution.

For some of the younger individuals, there is also the possibility that pressing

medical needs precluded dental care at and up to the time of survey completion (e.g., one or more cardiac conditions requiring surgery). For some older individuals, it is unclear if and what types of dental care were provided or recommended in, for example, prior dental visits.

Finally, participation in the TRIS project is voluntary and relies on recruitment via, for example, articles in newsletters and Facebook posts while the majority of literature on these groups is culled from hospital registries and/or national databases or based on case studies.^{1–3,15,16,18} The samples represented here may not be representative of the population (e.g., longevity).

Future research implications

While the authors cannot recommend specific dental treatment plans based on the results described here, it is clear that dental visits are essential within the cautions described here and in the literature concerning comorbid conditions of trisomy 18 and trisomy 13 (e.g., cardiac, respiratory, reflux). For example, the case study focusing on trisomy 13 notes, "Regular dental visits from an early age on should be encouraged to prevent extensive dental disease." (p. 95).¹⁶ The need for periodontal health is also emphasized across genetic conditions.²⁰ This draws attention to periodic examination for excessive plaque and related issues.

Further, oral health may not be considered a priority due to more pressing medical needs but should be part of ongoing medical supervision and follow up.²¹ With further investigation in this area, additional data can be examined to determine possible interactions of respiratory status and feeding methods, for example, with dental care. Primary feeding method and presence of reflux warrant further investigation into possible oral health and dental treatment needs. It would also be helpful to further examine interactions of dental care with phenotypic characteristics of cleft lip and/or palate, micrognathia and small mouth size.^{10,20,21}

Longitudinal data would provide additional insight into common oral

health needs and their treatment. The TRIS follow-up survey includes items on current respiratory conditions, feeding methods and several oral health needs and their treatment (excessive plaque, tooth decay and gum disease). Review and analyses of multiple years of data would contribute to greater understanding of oral health needs and assist to create a profile for treatment decisions. To this end, efforts are underway to select a sample of individuals with a minimum of three completed TRIS follow-up surveys (for a total of 4 years of data including initial completion of the TRIS Survey) and analyze resulting oral health data.

It is also important to implement recommendations highlighted in available studies and described here such as use of antibiotics prior to dental procedures in individuals with trisomy 18 and treatment in a dental office rather than a hospital setting to reduce the likelihood of infection.¹⁵ Other authors endorse the inclusion of dental professionals in the health care team for individuals with trisomy 18 over the age of one year emphasizing customized preventive programs.¹² It is imperative that dental care is included in care plans. The various specialists involved in overseeing management must be aware of oral health concerns and their potential impact on management such as amount and type of tube feeding and meeting respiratory needs.

Survey items did not ask about home care for primary and permanent teeth. This area is an additional avenue to investigate with individuals with trisomy 18 and trisomy 13. Delayed eruption of primary teeth and/or need for tube feeding should not preclude a home care regimen. Early and ongoing efforts to reduce plaque, for example, can have positive effects on oral health. Dentists should assist parents and caregivers with strategies for daily tooth brushing and related activities which meet the unique needs of individuals with trisomy 18 and trisomy 13 (e.g., high, arched palate, cleft lip and/or palate, micrognathia).

Conclusions

The sample described here offers initial results of routine and specialized dental care for individuals with trisomy 18 and trisomy 13. It is hoped this information will inform dental professionals in the provision of care for these unique groups.

Conflicts of interest

The authors have no conflicts of interest.

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References

- Irving C, Richmond S, Wren C, Longster C, Embleton ND. *Changes in fetal prevalence and outcome for trisomies 13 and 18: a population-based study over 23 years. J Matern Fetal Neo Med* 2011;24:137-41.
- Vendola C, Canfield M, Daiger SP, et al. Survival of Texas infants born with trisomies 21, 18 and 13. *Am J Med Genet Part A* 2010; 152A:360-6.
- Wu, JW, Springett, A, Morris, JK. Survival of trisomy 18 (Edwards Syndrome) and trisomy 13 (Patau Syndrome) in England and Wales: 2004–2011. *Am J Med Genet Part A* 2013;161A:2512-8.
- Petry P, Polli JB, Mattos VF, et al. Clinical features and prognosis of a sample of patients with trisomy 13 (Patau syndrome) from Brazil. *Am J Med Genet A* 2013;161A(6): 1278-83. doi: 10.1002/ajmg.a.35863.
- Bruns DA, Campbell E. Nine children over the age of one year with full trisomy 13: a case series describing medical conditions. *Am J Med Genet Part A* 2014b;164A(12): 2987-95. doi:10.1002/ajmg.a.36689.
- Bruns DA, Campbell E. Twenty-two long-term survivors with full trisomy 18: presenting and current medical conditions. *Am J Med Genet Part A* 2014a;164A(3): 610-19. doi:10.1002/ajmg.a.36318.
- Imataka G, Nitta A, Suzumura H, Watanabe H, Yamanouchi H, Arisaka O. Survival of trisomy 18 individuals in Japan. *Genet Counsel* 2007;18:303-8.
- Cereda A, Carey JC. The trisomy 18 syndrome. *Orphanet J Rare Dis* 2012;7(1):81. doi:10.1186/1750-1172-7-81.
- Carey JC. Trisomy 18 and trisomy 13 syndromes. In: Suzanne B. Cassidy, Judith E. Allanson, eds. *Management of Genetic Syndromes*. 3rd ed. Hoboken, NJ: Wiley-Blackwell; 2010:807-23.
- Bruns DA, Springer SA. Feeding changes in children with trisomy 18: longitudinal data on primary feeding method and reflux identification and treatment. *Top Clin Nur* 2013;28(4):324-34. doi:10.1097/01.TIN.0000437408.58941.d4.
- Jones K, Jones M, del Campo M. *Smith's Recognizable Patterns of Human Malformation*, 7th ed. Philadelphia: Elsevier Saunders; 2013: 14-19, 20-23.
- Ribeiro RR, dos Santos BM, Stuani AS, deFreitas AC, deQueiroz AM. Dental findings and dental care management in trisomy 18: case report of a 13-year-old "long-term survivor". *Spec Care Dentist* 2006;26:247-51. doi: 10.1111/j.1754-4505.2006.tb01662.x.
- Tatakis DN, Milledge TJ. Severe gingival recession in trisomy 18 primary dentition. A clinicopathologic case report of self-inflicted injury associated with mental retardation. *J Periodontol* 2000;71(7):1181-6.
- Smith A, Field B, Learoyd BM. Trisomy 18 at age 21 years. *Am J Med Genet Part A* 1989; 34(3):338-9.
- de Queiroz AM, Raffaini MSGG, deCamargo LM, et al. Orofacial findings and dental treatment in an 8-year-old patient with trisomy 18: a case report. *J Dent Childr* 2007;74(1):67-72.
- Klein U, Buehler BA. Dental management of an adolescent with trisomy 13 syndrome: a case report. *J Dent Childr* 2004;71(1):91-5.
- Butler MG, Hayes BG, Hathaway MM, Begleiter ML. Specific genetic diseases at risk for sedation/anesthesia complications. *Anesth Analg* 2000;91(4):837-55.
- Pollard RC, Beasley JM. Anaesthesia for patients with Trisomy 13 (Patau's syndrome). *Pediatr Anesth* 1996;6(2):151-3.
- Lin HY, Lin SP, Chen YJ, et al. Clinical characteristics and survival of trisomy 13 in a medical center in Taipei, 1985–2004. *Pediatr Int* 2007;49:380-6.
- Pizzo G, Re DL, Piscopo MR, Pizzo I, Giuliana G. Genetic disorders and periodontal health: a literature review. *Med Sci Monit* 2009;15(8):RA167-78.
- Carey J. Perspectives on the care and management of infants with trisomy 18 and trisomy 13: striving for balance. *Curr Opini Pediatr* 2012;24(6):672-8. doi:10.1097/MOP.0b013e3283595031.