

ORIGINAL ARTICLE

Assessment of orofacial characteristics and oral pathology associated with cri-du-chat syndrome

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OBJECTIVE: To obtain a deeper insight into the difficulties individuals with cri-du-chat syndrome experience by means of the analysis of the most common features and oral pathology observed in the subjects enrolled in the study.

SUBJECTS AND METHODS: Intra-oral and extra-oral features of a total of 33 patients with cri-du-chat syndrome (the larger sample so far analyzed) through their clinical and photographic examination. Models, orthopantomographies, and telerradiographies have been collected to establish a pattern as accurate as possible of the oral pathology associated with these patients.

RESULTS: The present descriptive study shows that patients with cri-du-chat syndrome present with a series of orofacial features such as mandibular retrognathism, high palate, and variable malocclusion, more commonly anterior open-bite. Most patients also present with perioral muscle relaxation with labial incompetence and short philtrum. As regards oral pathology, these patients suffer dental erosions provoked by gastroesophageal reflux and attritions because of intense day-and-night bruxism.

CONCLUSION: The odontologists' familiarity with the orofacial pathology associated with cri-du-chat syndrome and with the specific needs such disorder conveys should improve the quality of the buccodental treatment these professionals may offer to these patients.

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Keywords: cri-du-chat syndrome; oral pathology; dental pathology; periodontal pathology; periodontal microbiology

Introduction

Assessing and improving the quality of the dental care for a particular population involve the identification of

specific needs to adapt assistance to the given deficiencies. Among certain populations, odontological care and dental health practice are unsatisfactory (Douglass *et al*, 2004; Altun *et al*, 2010; Anders and Davis, 2010). Furthermore, this inadequate care is usually for those of a lower socioeconomic status and/or individuals with rare genetic disorders commonly associated with psychic and/or motor impairment. Those who have been diagnosed with cri-du-chat syndrome fall into this category.

Cri-du-chat syndrome is a chromosomal disorder characterized by severe intellectual and psychomotor development delay commonly associated with total or partial deletion of the end of the short (p) arm of chromosome 5. The most identifiable feature of this syndrome is a monotone weak catlike cry in infants, for which it is named (Lejeune *et al*, 1963). At present, this syndrome has an incidence of one in 37 000 newborns (Niebuhr, 1978), being slightly more prevalent among females (3:1), and it is not restricted to any ethnic group, geographical area, or prenatal association (Rodríguez-Caballero *et al*, 2010). Within the deleted chromosome region, we distinguish two main regions: band 5p15.3, which correlates with the characteristic catlike cry, and band 5p15.2 associated with dysmorphism, microcephaly, and mental retardation (Mainardi *et al*, 2001).

As of now, very little is known about the oral pathology associated with this syndrome. Few studies on this topic report features such as mandibular microretrognathism, anterior open-bite, high palate, and more rarely cleft palate (Scully and Davison, 1979). No further anomalies have been reported, as of yet, and the information about this syndrome is scarce.

In view of the lack of studies revising the dental condition of patients with cri-du-chat syndrome, we carried out the present study aiming to approach the specific needs and difficulties of these patients. To do so, we performed a descriptive analysis of the orofacial features and common oral pathology observed in the subjects enrolled in the study as well as their behavioral characteristics and other aspects as regards clinical management that may be relevant for the professional odontologist.

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Material and methods

We studied 33 patients with cri-du-chat syndrome belonging to the Asociación Nacional de Maullido de Gato (Cri du Chat Syndrome Spanish National Association) who were diagnosed in the School of Odontology of Seville from October 2008 through September 2009. As far as we know, this is the largest sample of this disorder so far reported.

The study was performed with the prior written consent of the parents or legal guardians of the patients and in compliance with the ethical principles for medical research involving humans as stated by the Helsinki Declaration of 2002. Likewise, all information regarding the patients was strictly confidential. The study was approved by the Ethical Committee of the University of Seville.

We recorded the medical history of the patients and carried out a complete clinical evaluation. The diagnosis of the syndrome was confirmed by means of a karyotype test.

The information collected included demographic and anthropometric data at birth and at later follow-up (birthweight, birth week, maternal lactancy, use of bottle feeding, and pacifier), major and minor malformations, and other medical problems. It was also registered information about general aspects related to prenatal incidents during pregnancy (toxemia, vaginal bleeding, maternal irradiation, and others).

Systemic anamnesis focused on the phenotypical features most commonly associated with this syndrome, more specifically, intellectual and motor development delay, muscular hypotonia and generalized hyperlaxity, aggressiveness and hyperactivity, sensory alterations such as hypoacusia or blindness, heart alterations, scoliosis, and gastrointestinal alterations.

The parents or guardians also completed several questionnaires on the oral health of the patients, oral habits, oral parafunctions, drooling, hygienic-dietetic habits, behavior, and odontological experience of patients prior to our study.

Following anamnesis, we carried out a thorough examination of patients. First, we analyzed the presence or absence of the following craniofacial characteristics: hypertelorism, epichantus, flat wide nasal bridge, short philtrum, down-turned lips, and low-set ears. We observed possible asymmetries, malformations, and scars in face and neck, facial profile, labial and perioral mucosa, and we performed a complete examination of the temporomandibular articulation.

Next, we examined and palpated the following soft tissues: lips, cheeks, tonsils, soft palate pillars, tongue, the floor of the mouth, hard and soft palate, frenum, and finally the gums. The pathological alterations observed were included in the clinical history of the patient.

Subsequently, we carried out a clinical examination of the teeth to count the number of existing teeth, dental defects (alterations in the number, color, shape, location, and morphology of the teeth), missing pieces, active and/or static decayed teeth, as well as the state of fillings and endodontics, if any.

Patients underwent photographic extra-oral and intra-oral examination (frontal, lateral, and middle third perspective; and frontal, lateral, and occlusal perspective, respectively) and radiographic analysis using orthopantomography and lateral radiography of the cranium, in those patients who could stay still long enough for the correct performance of the technique. The maxillary records of patients were taken to obtain the corresponding plaster models.

Not all examinations and questionnaires were able to be fully completed for all patients; therefore, the total number of patients for which data were recorded is specified in each section of the results.

The degree of disability was determined according to the official certificate issued by the competent regional authority. Guidelines for assessing the level of disability do not focus so much on the extent of the deficiency, but on its effect on the patient's ability to perform activities of daily living.

Current legislation recognizes five categories or levels of disabilities, sorted from the lowest to highest percentage: Grade 1 or no disability, 0%, Grade 2 or mild disability between 1% and 24%, Grade 3 or moderate disability between 25% and 49%, Grade 4 or severe disability between 50% and 70%, and Grade 5 or very severe disability, corresponds to individuals with 75% or more (Order in Council 1971/1999 Spain).

The degree of mental retardation was determined by the WHO criteria described in the ICD-10. According to which, we distinguish four degrees of mental retardation: mild mental retardation (IQ 50–69), moderate mental retardation (IQ 35–49), severe mental retardation (IQ 20–34), and profound mental retardation (IQ < 20) (World Health Organization, 2007).

An exploration of the skull was carried out, including the evaluation of the cranial perimeter. The diagnosis of microcephaly was reached based on a measure of cranial perimeter 3 DS below normal and pediatric reports of patients for which clinical documents were available.

Hyperlaxitud was diagnosed according to Brighton criteria [benign joint hypermobility syndrome (BJHS)] (Beighton *et al*, 2000). The BJHS is diagnosed in the presence two major criteria, or one major and two minor criteria, or four minor criteria. Two minor criteria will suffice where there is an unequivocally affected first-degree relative (Beighton *et al*, 2000).

Cavities and periodontal diseases were diagnosed following the guidelines established by the WHO (Boraz, 1990), as detailed later. We assessed the rate of decayed, missing teeth, and fillings (CAOD, total of teeth with caries or missing or with restoration divided by the number of teeth in the mouth), considering only permanent teeth with the exception of the third molar. Dental crowns counted as fillings when they resulted from decayed teeth and as healthy teeth when they acted as bridge abutments or were secondary to trauma.

Community periodontal index (CPI) was used to examine the periodontal condition of patients. Children below 11 usually presented with false pouches without the loss of insertion. Therefore and following the WHO recommendations (WHO, 1997), we only used codes 0,

1, and 2 in these patients (health, bleeding, and the presence of calculus).

The behavioral profile, the conduct, and the clinical management of the individual were evaluated across three scales of reference: Houpt Scale of Movement (Houpt *et al*, 1985), Classification of the Conduct according to the Frankl Scale (Frankl *et al*, 1962), and Scale for Clinical Management of the Patient (Bensberg *et al*, 1966).

The data were analyzed using SPSS 17.0 software for Windows (LEAD Technologies, International Business Machines Corp., Armonk, NY, USA). Univariate analysis of the results consisted of a descriptive analysis of the variables. Chi-square test was used to investigate the relationship between parameters.

Results

We studied a total of 33 patients, 13 males (39.39%) and 20 females (60.61%). Mean age of patients was 14.65 ± 10.19 years (interval 2–35), with a degree of disability of $73.42 \pm 15.99\%$ (Table 1).

The information gathered is shown in six tables. The Table 1 show the epidemiologic data corresponding to the neonatal period. Our patients had an average birthweight of 2382.65 ± 580.75 g and a gestational age of 38.75 ± 9.14 weeks.

The Tables 2 and 3 show the general features (Figure 1) and social background of patients. We observe that 34.37% of our patients brush their teeth once a day, usually before going to bed, and 31.25% three times a day (31.25%).

The Tables 4 and 5 show the buccodental pathology of patients. There is a high prevalence of mandibular microretrognathism. High palate is also found but in a lower proportion. As regards the CAOD of our patients, the value obtained was 0.28 ± 0.30 .

Table 6 focuses on the habits of patients with cri-du-chat syndrome; 75% of our patients showed euphoric, cheerful, and sociable personalities, whereas only a small number of them had a reserved and emotionally detached character. Considering the descriptive nature of our study, the following section is devoted to the detailed explanation and discussion of the different data

Table 1 Epidemiological data and information corresponding to the neonatal period of patients

Variables	Results			
Age (n = 32)	14.65 ± 10.19 years (interval 2:35)			
Sex (n = 33)	Male 13 (39.39%)		Female 20 (61.61%)	
Disability degree (n = 32)	73.42 ± 15.99%			
Mental handicap degree (n = 32)	Mild 1 (3.12%)	Moderate 16 (48.48%)	Severe 10 (31.25%)	Profound 5 (15.65%)
ASA (n = 32)	I 0 (0%)	II 31 (96.87%)	III 1 (3.12%)	IV 0 (0%)
Birthweight (n = 32)	2 383.65 ± 580.75 g			
Birth week (n = 32)	38.75 ± 9.14 weeks			
Suction at birth (n = 32)	Yes 9 (28.12%)		No 23 (71.87%)	
Suspicion diagnosis by typical catlike cry (n = 32)	Yes 30 (93.75%)		No 2 (6.25%)	

Table 2 General characteristics of patients under study

Variables	Result			
	Present (n)	Present (%)	Absent (n)	Absent (%)
Microcephaly (<i>n</i> = 32)	2	6.25	30	93.75
Hypertelorism (<i>n</i> = 32)	30	93.75	2	6.25
Epicanthus (<i>n</i> = 32)	30	93.75	2	6.25
Flat wide nasal bridge (<i>n</i> = 32)	30	93.75	2	6.25
Short philtrum (<i>n</i> = 32)	31	96.87	1	3.12
Low-set ears (<i>n</i> = 32)	25	78.12	7	21.87
Facial asymmetry (<i>n</i> = 32)	22	68.75	10	31.25
Divergent strabism (<i>n</i> = 31)	17	54.83	14	45.16
Hyperlaxity (<i>n</i> = 32)	25	78.12	7	21.85
Hypertony (<i>n</i> = 32)	5	15.62	27	84.37
Hypotony (<i>n</i> = 32)	28	87.5	11	34.37
Esophageal regurgitation (<i>n</i> = 32)	21	65.62	11	48.38
Dysphagia (<i>n</i> = 31)	16	51.61	15	48.32
Cardiac alterations (<i>n</i> = 29)	14	48.27	15	51.72
TMJ pathology (<i>n</i> = 13)	2	15.38	11	84.61

Variables	Results				
Social profile (<i>n</i> = 31)	Euphoric, sociable 23 (74.19%)		Autistic, antisocial 8 (25.80%)		
Hyperactivity (<i>n</i> = 32)	Yes 23 (71.87%)		No 9 (28.12%)		
Aggressiveness (<i>n</i> = 32)	Yes 21 (65.62%)		No 11 (34.37%)		
Behavior (<i>n</i> = 33)	1 11 (31.25%)	2 14 (43.75)	3 6 (18.75)	4 2 (6.25%)	
Movement (<i>n</i> = 32)	1 6 (18.75%)	2 13 (40.62%)	3 10 (31.25%)	4 3 (9.37%)	
Management (<i>n</i> = 30)	1 0 (0%)	2 2 (6.25%)	3 1 (3.12%)	4 4 (18.75%)	5 23 (71.87%)

Table 3 Social and behavioral profile and clinical management remarks in patients with cri-du-chat syndrome

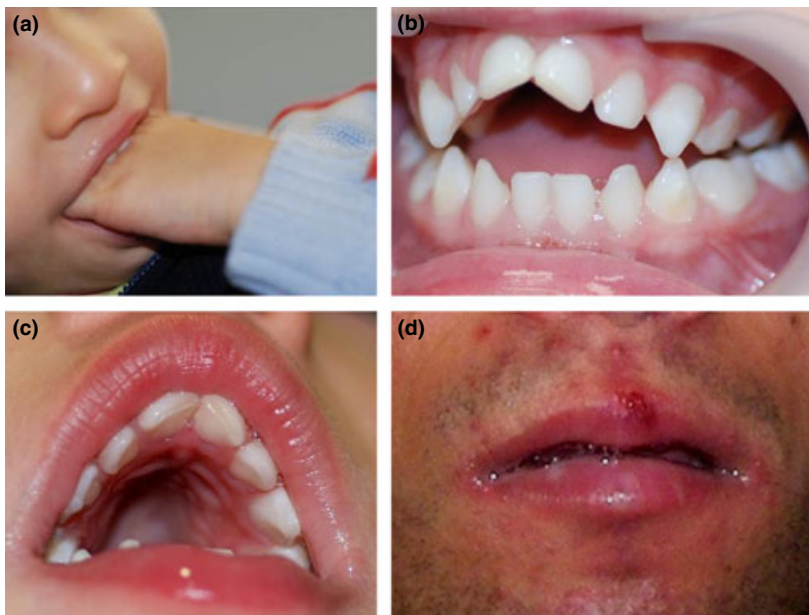


Figure 1 General features found in Cri-du-Chat patients. (a) Oral habit: hand suction. (b) Anterior open bite and posterior cross bite. (c) High palate. (d) Self-harm and dribbling

Table 4 Prevalence of buccodental alterations

Variables	Results	
	Present (%)	Absent (%)
Anterior open-bite(<i>n</i> = 30)	19 (63.33)	11 (36.66)
Mandibular retrognathia (<i>n</i> = 32)	29 (90.62)	3 (9.37)
High palate (<i>n</i> = 29)	16 (55.17)	13 (44.82)
Dental anomalies (<i>n</i> = 23)	13 (56.52)	10 (43.47)
Perioral muscle hypotonia (<i>n</i> = 32)	31 (96.87)	1 (3.12)

collected. Association between gender and mental handicap degree with buccodental alterations, CPI, and hygienic–dietetic habits of patients is shown in Table 7.

Discussion

This is a descriptive study whose sample of patients is the largest so far reported in the literature as regards the study of the oral pathology affecting patients with cri-du-chat syndrome. Although it was impossible to find a control group for comparison with our sample because of the heterogeneity of the items studied, the information collected provides a thorough picture of the

buccodental pathology and characteristics of patients with this syndrome. The discussion will proceed following the order of presentation of the different data collected.

Neonatal period

In 93.75% of our patients, suspicion diagnosis was established upon the presence of typical catlike cry at birth, whereas in the rest of patients, the disorder was diagnosed on account of the several clinical manifestations, being a cytogenetic analysis vital to establish a definite diagnosis.

Average birthweight was 2382.65 ± 580.75 g, and the gestational age approached normal values (38.75 ± 9.14 weeks). These data agree with previous reports in relation to the variables (Duarte *et al*, 2004).

Psychomotor development delay occurred in all patients under the study. Of them, 71.87% showed altered suction at birth (which prevented maternal lactancy), generalized muscular hypotonia (87.5%), dysphagia (51.61%), and gastroesophageal regurgitation (65.62%). Delayed development and growth during the first 2 years of life, to a large extent attributed to the difficulties these

Table 5 Community periodontal index (CPI)

	Health (%)	Bleeding (%)	Tartar (%)	Calculus 4-5 (%)	Calculus <5 (%)	Non-recorded (%)
CPI 1 (<i>n</i> = 32)	7 (21.87)	0 (0)	16 (50)	5 (15.62)	3 (9.37)	1 (3.12)
CPI 2 (<i>n</i> = 32)	8 (25)	1 (3.12)	19 (59.37)	4 (12.5)	0 (0)	0 (0)
CPI 3 (<i>n</i> = 32)	8 (25)	2 (6.25)	15 (46.87)	4 (12.5)	2 (6.25)	1 (3.12)
CPI 4 (<i>n</i> = 32)	8 (25)	2 (6.25)	16 (50)	5 (15.62)	0 (0)	1 (3.12)
CPI 5 (<i>n</i> = 32)	7 (21.87)	1 (3.12)	20 (62.5)	4 (12.5)	0 (0)	0 (0)
CPI 6 (<i>n</i> = 32)	6 (18.75)	2 (6.25)	15 (46.87)	6 (18.75)	3 (9.37)	0 (0)
Total CPI (<i>n</i> = 192)	44 (22.9)	8 (4.16)	101 (52.60)	28 (14.58)	8 (4.16)	3 (1.56)

Table 6 Hygienic–dietetic habits of patients

Variable	Results		
Daily teeth brushing frequency (<i>n</i> = 32)	1 every 2 days 3 (9.37%)	1 11 (34.37%)	2 8 (25%) 3 10 (31.25%)
Who does the brushing? (<i>n</i> = 31)	Child 18 (56.06%)	Father, mother or tutor 6 (19.35%)	Both 7 (22.58%)
Diet type (<i>n</i> = 31)	Soft 9 (29.03%)	Regular 22 (70.96%)	

Table 7 Association between gender and mental handicap degree with study variables (chi-square test)

	Gender	Mental handicap degree
CPI	0.002*	0.000*
Daily teeth brushing frequency	0.5	0.494
Who does the brushing?	0.731	0.923
Diet type	0.675	0.098
Mandibular retrognathia	0.159	0.545
High palate	0.436	0.304
Anterior open-bite	0.592	0.701
Dental anomalies	0.179	0.515
Perioral muscle hypotonia	0.431	0.518

CPI, Community periodontal index.
 *Statistically significant at *P* < 0.05.

patients have to eat, has been described by other authors (Cornish and Bramble, 2002; Mainardi *et al*, 2006).

Hygienic and dietetic habits

No previous studies have focused on the hygienic or dietetic habits of these patients. In our study, we observe that most patients brush their teeth once a day (34.37%), usually before going to bed, followed by those who brushed their teeth three times a day (31.25%). Over 50% of them brushed their teeth by themselves without being controlled by an adult (58.06%). However, several studies claim the necessity of adult supervision to ensure correct teeth brushing in patients with psychic impairment. This control normally takes place until the patients reach the age of 14, although very often, it could be necessary throughout the adulthood of these patients as well (Douglass *et al*, 2004).

It is essential to emphasize that in the relationship between the syndrome and the patient's oral hygiene, there are many confounding factors that prevent us from establishing a clear association, such as the type of

diet, the ability to understand, and the patient's ability to brush.

Craniofacial characteristics

As regards craniofacial characteristics, we observe epicanthus and hyperthelorum in most subjects (93.75%). Some authors consider these features as subjective, because of a visual effect the flat wide nasal bridge provokes (93.75%). According to Nieburh, it would be more precise to talk about 'eyes slightly separated' (Niebuhr, 1978), and he agrees with Sedano and Howard when he states that true hypertelorism is a rare characteristic of cri-du-chat syndrome (Sedano *et al*, 1971). Other common features, also reported in previous studies, were short philtrum (96.87%), low-set ears (78.12%), facial asymmetry (68.75%), strabismus (54.83%), and convex profile with maxillary mandibular retrognathism (93.54%). Conversely, microcephaly (6.25%) and a full-moon face (25%), both features being associated with cri-du-chat syndrome, were only observed in a reduced number of our patients. This may be due to the fact that subjects presenting with those two characteristics are very young (in their early years of life), whereas the average age of the patients included in our sample were over 14.

Oral pathology

Our study showed a high prevalence of mandibular retrognathism (90.62%) and a lower rate of high palate (55.17%), without cleft palate in any of the subjects. Anterior open-bite (63.33%) seems to be, together with mandibular retrognathia, the most common malocclusive alteration in cri-du-chat syndrome. We also observe hypotonic perioral muscles (96.87%) and dental anomalies (56.72%) such as enamel hypoplasias 54%, dental opacities 33%, tooth agenesis 9%, supernumerary tooth 3%, root resorption 3%, macrodontia 6%, and transposed teeth 3%.

Temporomandibular articulation could be examined in 39.39% of patients, 15.38% of whom showed clicks, crackling, pain, and/or mandibular deviation during mouth opening. These data match that reported by other authors, although our study is more exhaustive (Orudugba and Akindavomi, 2008; Altun *et al*, 2010).

CAOD index

Generally, the prevalence of untreated cavities in developed countries remains very high, especially among less favored and marginal population groups (Holt *et al*, 1996; Truin *et al*, 1998).

The objectives set by the WHO, with regard to buccodental health for the year 2000, have been met only in certain areas, resulting in a 50% reduction in cavities among children aged 5–6 and achieving a CAOD below three in 12-year-old boys (OMS (Organización Mundial de la Salud), 1984). Mean CAOD observed in our study was 0.28 ± 0.30 , which is considered a low index. We found a proportional increase in CAOD index with age, mainly because of the higher number of missing teeth either by trauma, periodontal disease, or by cavities. Such index, as well as the next, has not been reported by other authors studying cri-du-chat syndrome.

CPI index

A reduced number of our patients showed a healthy periodontal condition (22.9%), and more than half of them showed tartar in one or more quadrants (52.69%). We found 4- to 5-mm pouches in young subjects (14.58%) and pouches larger than 5 mm in three of the patients being studied (4.16%).

Behavioral patterns and odontological management

Of our patients, 77.8% showed euphoric, cheerful, and sociable personalities, whereas only a reduced number of them had a reserved and emotionally detached character. Prior studies describe most of these children as gentle and affectionate (Mainardi *et al*, 2006) and only exceptionally do we find patients with such characteristics as avoidance or social isolation, typical of autism (Altun *et al*, 2010).

According to the Frankl behavior rating scale (Table 3), most of the patients being studied showed a definitely negative (31.25%) and negative behavior (43.75%), whereas a reduced number showed a positive (18.75%) and definitely positive behavior (6.25%).

Our study shows that 90.2% of the patients required treatment under deep sedation or general anesthesia, and only a small number of them could be treated using movement restriction techniques.

It is important to note the contrast observed between the behavior rating scale, according to which 75% of the patients had negative or definitely negative behavior, and the 90.62% of patients who did not permit ambulatory odontologic treatment. This is explained by the influence of other factors that determine the final decision: systemic pathology of patients, the number of interventions they would have to undergo in case of being treated, the specific personal circumstances of each

patient, and once all the options have been presented, the criteria of parents, or legal guardians of the patient (Liu *et al*, 2010).

Conclusion

Our descriptive study shows that patients with cri-du-chat syndrome present with such orofacial features as mandibular retrognathism, high palate, and variable malocclusion, more commonly anterior open-bite. Most of them present with perioral muscle hypotonia with labial incompetence and short philtrum.

As regards oral pathology, these patients present with excessive dental erosions provoked by gastroesophageal reflux and attritions owing to intense day-and-night bruxism. These patients also show dental alterations as regards structure, size, position, and number of pieces; and more precisely enamel hypoplasia, opacities, and agenesis. Oral hygiene is quite poor in these patients who also show a significant number of active cavities and gingivitis and/or periodontal disease.

As regards their social and behavioral patterns, they usually have sociable, euphoric, and cheerful personalities. Yet, in most of them, hyperactivity, aggressiveness, and continuous uncontrolled movements will complicate ambulatory odontological treatment. As a result, a high percentage of them will require general anesthesia.

Unawareness of the specific buccodental problems affecting these patients is surely an obstacle to their adequate access to oral health. The greater the knowledge the professional odontologist has about orofacial pathology associated with cri-du-chat syndrome and the specific needs of these patients, the better the quality of buccodental health assistance he can offer.

Author contributions

Ángela Rodríguez-Caballero contributed to conception and design, data acquisition, analysis and interpretation of data. Daniel Torres-Lagares contributed to conception and design, analysis and interpretation of data. Rosa-María Yáñez-Vico contributed to drafting the article and to analysis of data. José-Luis Gutierrez Pérez contributed to interpretation of data and to drafting the article and critical review. Guillermo Machuca-Portillo contributed to drafting the article and critical review and to final approval of the version to be published.

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