

# Cherubism: Clinicoradiographic Features and Treatment

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## ABSTRACT

**Objectives:** Cherubism is a congenital childhood disease of autosomal dominant inheritance. This disease is characterized by painless bilateral enlargement of the jaws, in which bone is replaced with fibrous tissue. The condition has *sui generis* clinical, radiographic and histological features, of which the clinician should be aware for a better differential diagnosis in the presence of a fibro-osseous lesion affecting the bones of the maxillomandibular complex. The purpose of present paper was to review the literature and to report the most important aspects of cherubism in order to facilitate the study of this disease.

**Material and Methods:** Literature was reviewed about cherubism, emphasizing the relevant clinicoradiographic features and treatment. Literature was selected through a search of PubMed and Scielo electronic databases. The keywords used for search were adolescent, cherubism, cherubism/physiopathology, cherubism/treatment, cherubism/radiography. A manual search of the reference lists of the identified articles and the authors' article files and recent reviews was conducted to identify additional publications. Those studies that described new features about cherubism were included in this review.

**Results:** In total 44 literature sources were obtained and reviewed. Studies that described new features about cherubism physiopathology, diagnostics and treatment were reviewed.

**Conclusions:** Despite the exceptions, cherubism is a clinically well-characterized disease. In cases of a suspicion of cherubism, radiographic examination is essential since the clinical presentation, the location and distribution of the lesions may define the diagnosis. Histopathological examination is complementary. Nowadays, genetic tests should be used for final diagnosis of cherubism.

**Keywords:** adolescent; jaw diseases; cherubism.

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## INTRODUCTION

Cherubism is a rare disease of autosomal dominant inheritance characterized by painless, frequently symmetrical, enlargement of the jaws as a result of the replacement of bone with fibrous tissue [1-16,20,29]. The disease is also called familial fibrous dysplasia of the jaws, but recent genetic investigation has shown it to be a separate entity at the molecular level [7]. Furthermore Lannon et al. [30] mentioned necessity to distinguish cherubism from central giant cell granuloma and giant cell tumour of the jaws, with which it holds a false synonymy.

A molecular pathogenesis of cherubism has been proposed, with the detection of a mutation in the gene encoding SH3 - binding protein 2 (SH3BP2) [6,10,31] and possible degradation of the Msx-1 gene which is involved in the regulation of mesenchymal interaction during craniofacial morphogenesis [11]. It is believed that the different clinical manifestations of cherubism are due to the changes secondary to mutations or incomplete penetrance [10].

Cherubism is usually diagnosed in children aged 2 to 7 years, with the observation of exacerbation of its manifestations within the first 2 years after diagnosis [5,32] and of stabilization or even regression after puberty [1-14,20]. Boys are more affected than girls at the proportion of 2 : 1 [3-6,14,33].

Clinically, cherubism is characterized by bilateral enlargement of the mandible and/or maxilla, causing a rounded face and swollen cheeks accompanied by upward-looking eyes. This condition gives the patient the appearance of cherubs depicted in baroque artwork [1,4-12,14,16,20,34]; hence, the name of the disease introduced by Jones [17], who published the first four cases affecting the same family.

The purpose of the present paper was to review the literature and to report the most important aspects of cherubism in order to facilitate the study of this disease.

## MATERIAL AND METHODS

Literature was reviewed about cherubism, emphasizing the relevant clinicroadiographic features, physiopathology and treatment. Literature was selected through a search of PubMed and Scielo electronic databases. The keywords used for search were adolescent, cherubism, cherubism/physiopathology, cherubism/treatment, cherubism/radiography. A manual search of the reference lists of the identified articles and the authors' article files and recent reviews was conducted to identify additional publications. Those

studies that described new features about cherubism physiopathology, diagnostics and treatment were included in this review.

## RESULTS

### Pathogenesis

Although rare, cherubism is part of the differential diagnosis of bone diseases affecting the maxillomandibular complex and the professionals should therefore have knowledge of the disease. The most accepted theory regarding the pathogenesis of cherubism is its association with an autosomal dominant gene, i.e., family inheritance [1-9,11-17,23,35]. However, there are reports of the cases in which no criteria of heredity could be established [1-3,9,11,12,15,16,24], or in which an autosomal recessive pattern of inheritance was suggested [5]. In addition to genetic factors, Caballero and Vinals [14] indicated other possible causes of cherubism such as mesenchymal alterations during jaw development, an odontogenic origin or even hormonal and traumatic factors. Today we know that a mutation in the gene encoding SH3BP2 plays a role in the disease [6,7,10,11,21]. There are indications that the gene SH3BP2 plays a role in regulating the increased osteoblast and osteoclast activities that are seen in normal tooth eruption, and point mutations in the gene could cause pathologic activation of osteoclasts [21,31,36]. Silva et al. [11] reported the degradation of Msx-1 gene, which is involved in the regulation of mesenchymal interaction during craniofacial morphogenesis. In contrast, Sarda et al. [10] detected a point mutation at the SH3 binding site of the SH3BP2 gene, and Li and Yu [6] observed mutations in exon 9 of the same gene.

According to Hyckel et al. [26] cherubism is a location-stable phenomenon found only in the jaws with multiple occurrences. Furthermore, structure-associated process is a very likely link to the pathogenic mechanism. The authors defined cherubism as a genetically determined alteration of tooth germ development. They proposed the molecular model of cherubism pathogenesis which is based on interaction between a disturbed (due to mutation in SH3BP2) parathyroid hormone related protein (PTHrP) receptor with the Hox gene Msx-1 activity. Thus the temporal and spatial termination of the clinical symptoms is explained by SH3BP2 - dependent signal transduction pathways interfering with jaw morphogenesis. The cap stage of the second and third molars, a spatial compartmentation does not take place, being necessary for normal dental development. This leads to the dysregulation of mesenchymal

bone formation, and to the development of giant cell granulomas containing osteoclasts.

### Clinical characteristics and symptoms

Patients with cherubism, generally males at a proportion of 2 : 1 [2,3,5,7,14], present the same clinical characteristics: enlarged face due to swelling of the jaws which is bilateral in most cases, bone consistency of the lesion, intact mucosa, dental malocclusion, upward-looking eyes in the case of maxillary involvement, and absence of pain [2,4-9,11,12,14-17,37]. The concomitant presence of cervical and/or submandibular lymphadenopathy has been reported [8,13,14,16,23]. The first signs of manifestation of the disease are generally observed at about 2 years of age, followed by accelerated growth from 8 to 9 years and spontaneous interruption after puberty [1-3,5,9,11,12,14-16,24,29,38]. However, Ashraf [1] and Kalantar [4] suggested that, as observed from 3 to 7 years of age, growth might be more active after puberty. In addition, the age at recognition of symptoms varies according to the severity of the disease and degree of deformity. In such cases where the onset of symptoms of cherubism occurs after puberty, the time of remission of the process is expected to be prolonged [1,3,4]. In cases where the clinical symptoms are typical to cherubism, the final diagnosis should be based on radiographic findings and histology, because the clinical picture of cherubism at initial examination are similar to other lesions that cause mandible enlargement, such as autosomal dominant osteosclerosis [22].

### Dental alterations

Dental alterations associated with cherubism include early exfoliation of deciduous teeth, impaction and/or displacement of teeth [1,4-14,16,20], which radiographically seem to float in radiolucent areas, conferring the so-called "floating tooth appearance" [14,20]. In addition, ectopic tooth eruption, agenesis of permanent teeth, mainly of the second and third molars [26], due to involution of their germs [14], and root resorption of existing teeth are observed [4,5]. These alterations result in malocclusion as well as in problems of phonation and swallowing, the latter being exacerbated by flattening or inversion of the palatal cleft [1,4-6,8-12,14,16].

### Biochemical parameters

With regard to biochemical parameters, serum calcium and phosphorus concentrations and TSH, FSH, LH, T4 and T3 hormone levels are usually within normal limits,

but alkaline phosphatase levels might be elevated [1,5,8,10,11,13-15]. A constant finding in these patients is the enlargement of submandibular and cervical lymph nodes [5,8,13,14].

### Radiography

Radiographically, cherubism is characterized by expansive radiolucent, generally multiloculated lesions clearly delimited by cortical bone and distributed bilaterally in the posterior quadrants of the mandible and/or maxilla [6,8,9,12,20]. Bone alterations generally starts in the region of the angle and ascending ramus of the mandible, continue to the mandibular body, displacing the mandibular canal, and in some cases extending to the coronoid process [1-3,8,14,17,23]. Involvement of the condyle is rare [3,4,9,11]. In the maxilla, the injury process starts in the maxillary tuberosity region [4,5,9,10,14]. When process is starting in mandible, maxillary lesions manifest at a later time [14]. In more severe cases, infiltration of the orbital cavities may cause exacerbated exophthalmia and limiting ocular movements [1,11,15,16,39].

The lesions of cherubism can be classified according to their extent: grade I, bilateral involvement of the ascending ramus of mandible; grade II, bilateral involvement of the ascending ramus of mandible and maxillary tuberosity; grade III, complete involvement of the maxilla and mandible compromising the coronoid processes and condyles [1,4,16].

Radiographically identified multilocular radiolucent lesions mainly are affecting the body and ascending ramus of the mandible. The coronoid process might be involved [23,24] and the condition extends to the maxilla, preferentially to the region of maxillary tuberosity [20,23]. The radiolucencies are mainly bilateral and may affect the lower portion and fundus of the orbit which cortical bone appears to be thin but intact. The teeth are found to be displaced and impacted and root resorption is observed; the mandibular canal is often displaced [1-3,5,8,11,12,14-16,23]. The facial sinuses frequently appear to be obliterated, resuming their pneumatic function after regression of disease activity [5,14]. Some authors have reported involvement of the condyles [1,3,4].

### Diagnostics

Observed radiographic appearance might be confused with other lesions containing giant cells: hyperparathyroidism [5,8,9,18], and/or osteomalacia [19]. Regarding the differential diagnosis, it should be emphasized that, whereas central giant cell lesions affect the central portion of the mandibular body and giant cell

tumours rarely involve bone of the maxillomandibular complex, in cherubism the lesions are generally bilateral and involve both the maxilla and the mandible [3,18]. The radiographic techniques used for the diagnosis of suspected cases of cherubism include posteroanterior radiography of the jaws [5,16], orthopantomography [1-4,6-9,11,12,14] and teleradiography [3,14,16]. Computed tomography is a useful tool for the assessment of the damage caused by the process either during the analysis of disease progression or during surgical planning [3,5,7-9,40]. Advancements in virtual three-dimensional reconstruction of anatomic structures based on computed tomography or cone beam computed tomography data can provide for more predictable individual treatment planning [41]. According to Mnari et al. [42] magnetic resonance imaging is useful for identifying orbital involvement.

Similarity with central giant cell lesions and hyperparathyroidism is also present in the histological findings. As observed in these conditions, in cherubism multinucleated giant cells are found scattered throughout collagenous and vascularized fibrous connective tissue with the observation of dense connective tissue which might be highly or poorly cellularized and contain a smaller or larger number of collagen fibers, respectively, depending on the progression of the lesion [1,3-8,11,14]. This connective tissue is richly vascularised and mainly located around giant cells, with the possible presence of bone spicules and hemosiderin deposits [1,3-5,7,8,11-16,18]. It is believed that these giant cells have osteoclast activity [5,13]. Southgate et al. [13] found that multinucleated cells in cherubic lesions had phenotypic characteristics of osteoclasts, resorbing bone *in vitro*. The specific histological finding of cherubism is the presence of eosinophilic perivascular cuffing which, however, is not always present [3,14,24] and histology is of limited diagnostic significance [26]. Figures 1 to 4 shows the extraoral clinical and radiographic features of two boys (7 and 9 years old) with cherubism involving the angle and ascending ramus of the mandible. The cases were retrieved from the archives of the Discipline of Stomatology, São José dos Campos Dental School, UNESP. Figures 5 and 6 refer to the follow-up of the case shown in Figures 3 and 4 at nine years after diagnosis.

Laboratory tests contribute to the characterization of disease since hematological parameters such as calcium and phosphorus are normal and only alkaline phosphatase levels might be elevated [1-3,5,6,8,11,13,15,23]. According to George et al. [3], the increase in alkaline phosphatase can be explained by the phase of development in most individuals affected by the disease. In contrast, in hyperparathyroidism serum calcium and PTH levels are increased and phosphorus

levels are found to be normal or reduced [15,19].

Cytogenetic and molecular studies have been used on cherubism diagnostic, such as fluorescence *in situ* hybridization and quantitative analysis of Msx-1 expression in different tissues [26].

## Treatment

Once the diagnosis is established, therapeutic management should be evaluated. Treatment options include waiting for stabilization and spontaneous remission of the disease, tooth extraction in areas showing fibrous alterations, cosmetic osteoplasty of the affected jaws after regression of disease activity or, in the case of functional impairment, curettage of the lesions and treatment with calcitonin [32,34,43,44]. The policy of waiting for disease regression, followed by the evaluation of physiological bone remodelling, is the most recommended [3,5-8,11,12,14-16,23,26]. However, it is still uncertain whether this approach is the most effective one since only a few cases of long-term follow-up have been reported and in most cases submitted to curettage or jaw osteoplasty surgery was indicated early [1,3-5,17]. Curettage has been suggested to be as a good approach since this intervention stimulates bone replacement [3]. However, in his original study Jones [17] treated two boys by bilateral curettage and one girl was submitted to curettage on one side and multiple tooth extractions on the other side, with no difference in outcome being observed between the two sides. According to the author, this finding might indicate that cherubism is associated with odontogenesis and multiple extractions would resolve the case. There is general agreement that in extreme cases in which important functions are impaired, surgical intervention should be performed as early as possible [4,5,8,11,14,19]. Radiation therapy has been abandoned as a treatment of cherubism because of the potential risk of osteoradionecrosis or even malignant transformation of the process resulting in osteosarcoma [2,5,8,10,27]. When possible, follow-up is always a valuable choice [26].

According to Novack and Faccio [27], hypothesis that cherubism is caused by enhanced cytokine tumour necrosis factor  $\alpha$  (TNF- $\alpha$ ) production by myeloid cells due to an activating mutation in Sh3bp2 not only represents a major advancement in the understanding of the disease but suggests new potential options for its treatment. Although cherubism was described more than 70 years ago, the rather sparse literature in this area has provided little insight on disease effective therapies. As a major pathogenic factor of the identification of TNF- $\alpha$  for the patients with this rare disease, is significant news. Anti-TNF therapies are already in clinical practice for





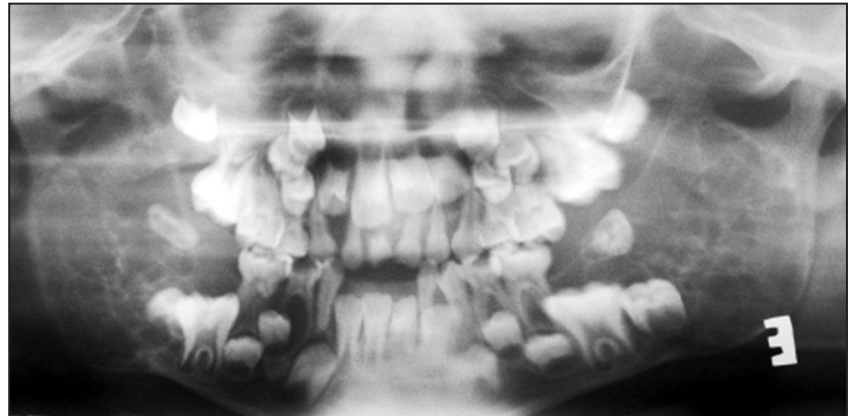
**Figure 1.** Photograph of a 7 year old boy with cherubism showing bilateral swelling at the mandibular angles.



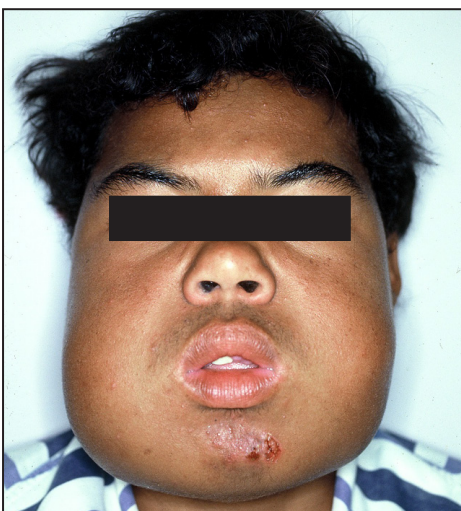
**Figure 2.** Orthopantomograph of the same patient as in Figure 1. Note the bilateral swelling caused by expansion related to multilocular bone lesions of the angle and ascending ramus of the mandible and coronoid process.



**Figure 3.** Photograph of a 9 year old boy with cherubism showing bilateral swelling of the angle and ascending ramus of the mandible, stretching the skin in the nasogenial region.



**Figure 4.** Orthopantomograph of the same patient as in Figure 3. The image reveals bilateral swelling of the angle, ascending ramus of the mandible and coronoid process caused by multilocular bone lesions. Note the lack of involvement of the condyles.



**Figure 5.** Photograph of the same patient as in Figure 3 after 9 years of follow-up, demonstrating expressive progression of the disease.



**Figure 6.** Orthopantomograph of the same patient as in Figure 5, revealing radiographic alterations characterized by greater radiopacity resulting from bone deposition and confirming the lack of involvement of the condylar regions. Note the malpositioning and retention of teeth.

the treatment of rheumatoid arthritis. If these drugs will prove the effectiveness in the treatment of cherubism it can be hoped that the interval from laboratory discovery to clinical use would be short [27].

A study by von Wowern [25] followed-up 18 patients over the long term and found a significant relationship between the grade of cherubism and maximal buccal bone expansion, as well as the age for normalization, and the grade of cherubism and number of missing teeth. Neither patient showed any radiolucencies or radiographic signs of sclerotic areas or sclerotic thickening of the mandibular ramus. In the same study, no larger surgical corrections with removal of the main part of the lesional tissue were performed in any of the patients.

A prosthetic treatment for a cherubism patient was reported by Yilmaz et al. [28]. The patient's physical appearance showed characteristic cherubic features. The maxillary canines, the second and third molars were missing and almost all mandible teeth were extracted before the patient was referred for treatment. Authors were concerned that implant placement into porous bone might result in lack of osseointegration and lead to complication. Thus, treatment plan consisted of fixed partial dentures preparation for the maxilla and an overdenture with copings for the mandible. The patient remained satisfied with the function and aesthetic result at the one year recall appointment.

## CONCLUSIONS

Despite the exceptions, cherubism is a clinically well-characterized disease which confers to the patient the appearance of a baroque cherub; therefore, this derived the name of the disease. In cases of a suspicion of cherubism, radiographic examination is essential since the clinical presentation and the location and distribution of the lesions may define the diagnosis. Histopathological examination is complementary. Nowadays, genetic tests should be used for final diagnosis of cherubism.

Knowledge of the clinical and radiographic alterations observed in patients with cherubism is important since the dentist might be the first professional sought for a diagnosis of this disease.

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

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