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Histology of alveolar bone and primary tooth roots in a case of cleidocranial dysplasia

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SUMMARY

Cleidocranial dysplasia is commonly reported as an autosomal dominant inherited condition with defective formation of clavicles, malformation of the craniofacial bones, very slow exfoliation of the primary teeth and failure of the eruption of the permanent dentition.
Lack of clinical resorption of the roots of the deciduous teeth and/or surrounding bone, lead to eruption failure of permanent teeth.
Histopathological study (light and scanning electron microscopy), in a case of cleidocranial dysplasia, gives prominence to the hypothesis of abnormal remodelling of bone and cementum.

KEY WORDS:

Cleidocranial dysplasia, bone, cementum.

RÉSUMÉ

La dysplasie cleïdocrânienne est un syndrome transmis sur le mode autosomique dominant. La maladie se caractérise par des dysplasies des clavicules et des os crânio-faciaux, un retard de la perte des dents temporaires et des anomalies d'évolution des dents permanentes.
L'absence de résorption cliniquement décelable des racines des dents temporaires et/ou de l'os environnant, entraîne des anomalies d'éruption des dents permanentes.
L'étude, en microscopie photonique et microscopie électronique à balayage de l'os et du cément de dents temporaires, permet d'avancer l'hypothèse d'un remodelage anormal des tissus calcifiés dans les cas de dysplasie cleïdocrânienne.

MOTS CLEFS:

Dysplasie cleïdocrânienne, os, cément.

INTRODUCTION

Cleidocranial dysplasia is an autosomal dominant inherited condition with multiple and severe skeletal and dental abnormalities (Gorlin et al., 1975).

The most striking features are the defective formation or absence of clavicles, malformation of the craniofacial bones and the eruption failure of the dentition.

Although cleidocranial dysplasia is a generalized bone dysplasia, literature is often focused on dental problems.

The primary dentition is said to develop in a relative normal way (Eisen, 1953), although it is very slow in exfoliating (Monasky et al., 1983: Jensen and Kreiborg, 1990). The permanent dentition is severely disturbed with predisposition for multiple supernumerary teeth, failure of eruption, ectopia and abnormal tooth morphology especially involving the roots (Rushton, 1937; Kirson et al., 1982; Yamamoto et al., 1989; Jensen and Kreiborg, 1990).

The severely delayed or arrested eruption of permanent teeth could be ascribed to various factors: presence of multiple supernumerary teeth (Rushton, 1937), malformed roots with lack of cellular cementum (Rushton, 1956; Alderson, 1960), high density of the jaw bone (Rushton, 1937), failure of alveolar bone resorption (Hitchin and Fairley, 1974; Migliorisi and Blenkinsopp, 1980).

According to Rushton (1937) and Hitchin and Fairley (1974), the frequently reported abnormality of inadequate deformed spike-shaped roots, crown distortion and hypoplasia in areas of compression from adjacent teeth, strongly suggests a disturbance in bony remodelling, which acts upon the dentition development and delay the eruption.

In the developing child this could include a failure of resorption of the roots of the deciduous teeth. Histopathological studies of maxillo-facial calcified tissues involved in the disease, used only one methodological approach.

The purpose of the present investigation is to complete description of clinical features by microscopical observations concerning dental and periodontal structures, using three different observation techniques.

From these results, it could be possible to throw light on the reasons for non eruption of teeth in a case of cleidocranial dysplasia.

MATERIAL AND METHODS

The 11-year-old French girl discussed in this paper showed many of the characteristics identifiable with cleidocranial dysplasia.

She was of short stature with complete aplasia of the clavicles and associated funneling of the rib cage towards the thoracic inlet. Her pelvis was hypoplastic as were the carpal, metacarpal, tarsal, and metatarsal bones.

Her skull exhibited brachycephalic deformity with frontal and parietal bossing.

The maxillary and zygomatic bones were hypoplastic with widening of the pyriform aperture. The paranasal air sinuses were poorly pneumatised, the palate was high vaulted but there was no cleft. Dento-alveolar abnormalities present were consistent with the syndrom, namely retention of the deciduous dentition, delayed eruption of the permanent dentition except the first permanent molars and lower central incisors, and the presence of mandibular supernumerary teeth (Fig. 1).

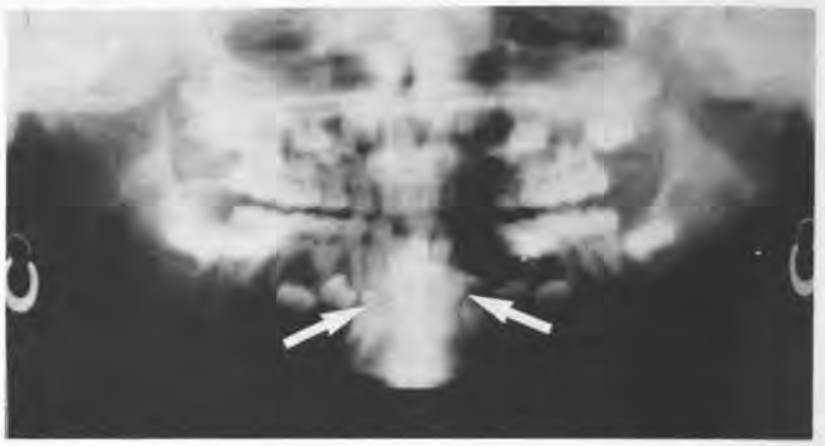

Fig. 1.: oro-dental panoramic radiography before surgical management. Arrows = supernumerary teeth.
Fig. 1.: radiographie panoramique dentaire pré-opératoire. Flêches = dents surnuméraires.

Radiographically the deciduous teeth showed little in the way of root resorption; the supernumeraries lay, between and apical to the deciduous roots, displacing the permanent incisors and canines towards the lower border of the mandible.

The supernumerary teeth were irregular in form, the morphology of the permanent teeth appeared quite normal with reduced root formation consistent with unerupted teeth.

The surgical intention in the management of this patient was to effect the eruption of the permanent dentition by removal of all the obstacles from their path. This was carried out first in the mandible. The operative technique entailed extraction of the still almost complete deciduous dentition, followed by the surgical exposure from the oral aspect of the alveolar bone of supernumerary teeth which were removed.

Having removed the dento-alveolar obstruction with preserving the plates of alveolus, normally permanent teeth should have made their eruption in a few months.

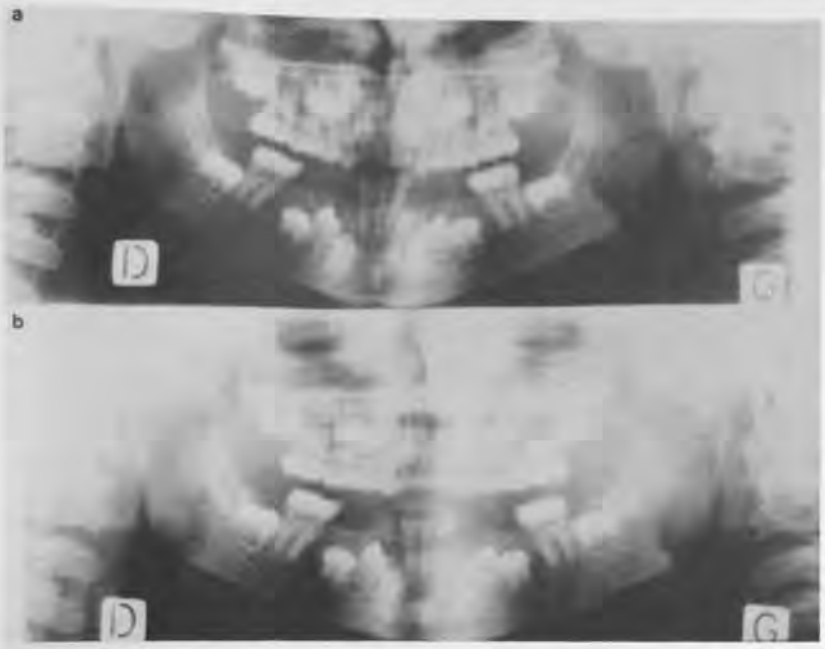

Fig. 2.: a - oro-dental panoramic radiography immediately after surgical exposure, and...
b - ...six months later.
Fig. 2.: a - radiographie panoramique dentaire post-opératoire, et...
b - ...six mois plus tard.

Six months later, we can observe absence of eruption of the mandibular permanent teeth (Fig. 2).

During surgical management, three deciduous molars clinically free of root resorption, and three bone fragments were removed and immediately immersed in a fixative solution (neutral formalin 4%). Undecalcified samples were then dehydrated in gradual ethanol solutions (from 70° to absolue ethanol), before inclusion in methyl-methacrylate resin (Prolabo).

Histological sections, 80 micrometers thickness, were obtained by microtomy technique (Isomet, Buehler LTD), and observed by light transmitted microscopy (Olympus BH-2).

Microradiograph were performed using an X-rays generator, with copper target (Philips), and observed by light transmitted microscopy (Olympus BH-2). Calibration of the projected image with aluminium wedges was not used.

Two bone fragments removed from alveolar crest were immersed in ethanol 70° as a fixative and then prepared for scanning electron microscopy observations (Hitachi S-450): removal of organic materials using sodium hypochlorite, dehydratation in ethanol solution (70°), use of critical point drying (CPD 010, Balzers Union) and coating by gold (EMscope).

RESULTS

Lack of cementum or presence of acellular cementum, surrounding dentin (Fig. 3) with regular tubuli arrangement (Fig. 4), predominates along roots on deciduous teeth (Fig. 3). Resorption areas appear, in this place (Fig. 4), and on apex (Fig. 5) subsequently filled by cellular cementum which is also present within the inter-radicular space.

Fig. 3.: d = dentin; ac = acellular cementum. Scale/1 cm = 1 mm.
Fig. 3.: d = dentine; ac = cément acellulaire. Echelle/1 cm = 1 mm.

Using micro-radiography, we note the presence of a voluminous globular calcification, and numerous micro-calcifications within the radicular dental pulp tissue (Fig. 6).

A great number of needle-shaped micro-calcifications is also present in periodontal ligament, far away from bone and cementum (Fig. 7).

The radio-density of these heterotopic mineralized structures is generally quite similar to those of bone or dentin (Fig. 6, Fig. 7).

By scanning electron microscopy, bone structure appears normal, without resorption lacunae. It exist a randomised distribution of osteocytic lacunae (osteoplsts) around nutritional canal (Fig. 8).

Light microscopy of this tissue, evidences resorption areas subsequently filled by new haversian bone (Fig. 9).

DISCUSSION

Our personal case report is in agreement with previous clinical descriptions reported by numerous authors from 1890 (Marie and Sainton, 1890) to our days (Monasky et al., 1983; Kalliala and Taskinen, 1962; Hasler and Vandermer, 1974; Järvinen, 1980).

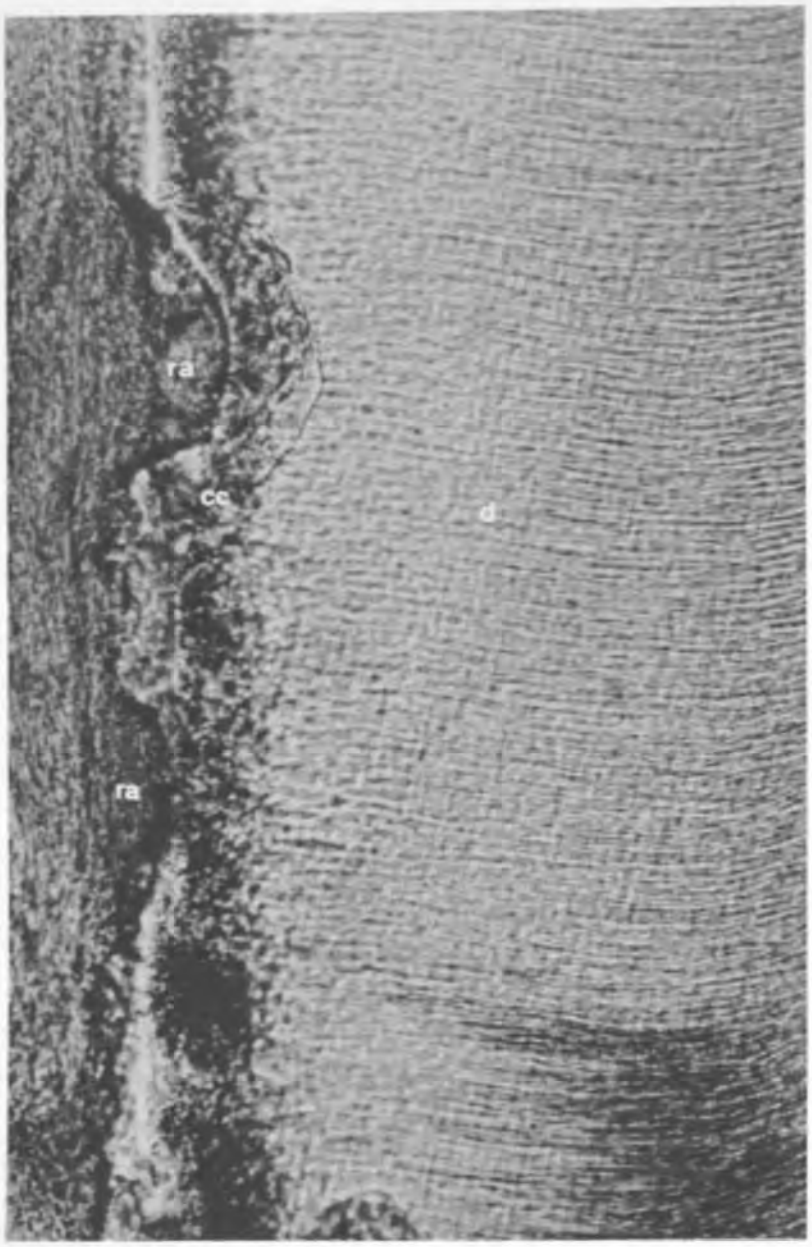

Fig. 4.: d=dentin; ra=resorption area; cc=cellular cementum. Scale/1 cm=70 µm.
Fig. 4.: d=dentine; ra=zone de résorption; cc=cément cellulaire. Echelle/1 cm=70 µm.

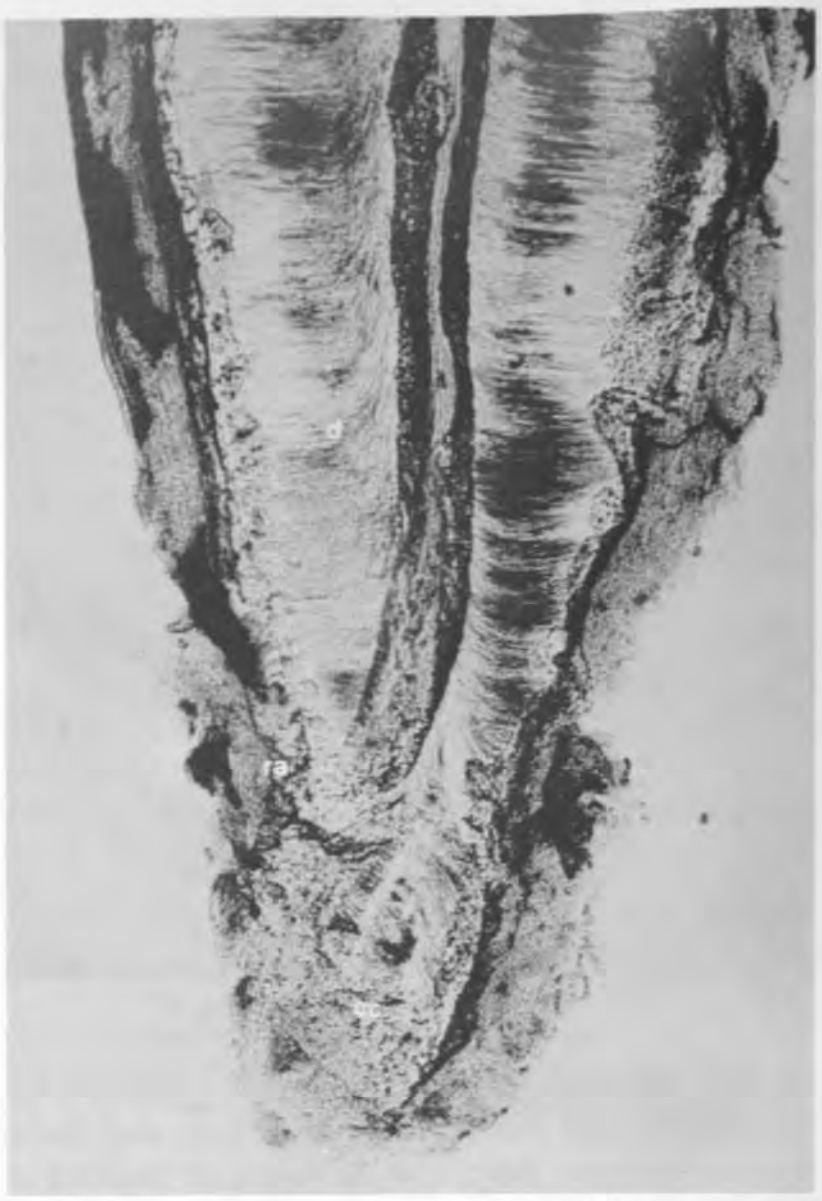

Fig. 5.: d=dentin; ra=resorption area; cc=cellular cementum. Scale/1 cm=0,35 mm.
Fig. 5.: d=dentine; ra=zone de résorption; cc=cément cellulaire. Echelle/1 cm=0,35 mm.

According to Migliorisi and Blenkinsopp (1980), and to Hall and Hyland (1978), the removal of bone upon permanent teeth facilitates their eruption. Our clinical findings are in contradictory with these features, and in agreement with those of Magnus and Sands (1974).

For this reason the understanding of the failure of shedding (deciduous teeth) or/and eruption (permanent teeth), in cleidocranial dysplasia, is of great interest.

The finding of arrested eruption, even in region without temporary teeth, supports the hypothesis of abnormal remodeling of calcified tissues, essentially bone and cementum (Jensen and Kreiborg, 1990).

In our case report, the non-eruption of two permanent incisors, canines and premolars, six months after surgical management of deciduous teeth and bone, favors the same idea.

Further evidence for this hypothesis is obtained from histopathological examination of the alveolar bone removed from patients with the disease.

Sections of bone supporting deciduous teeth (Hitchin and Fairley, 1974), and further sections from the bone supporting the unerupted molars in the case described by Migliorisi and Blenkinsopp (1980), showed abnormally dense trabeculation with mutiple reversal lines, indicating incomplete resorption phases.

Our histopathological study (SEM) reveals ultrastructural normal aspect of bone; but, light microscopic study shows, that when resorption lacunae exist, they seem systematically filled by bone apposition, indicating disturbances in the process.

The absence of cellular cementum, as originally reported by Rushton (1956) in permanent teeth, is difficult to elucidate. This author studied histologi-

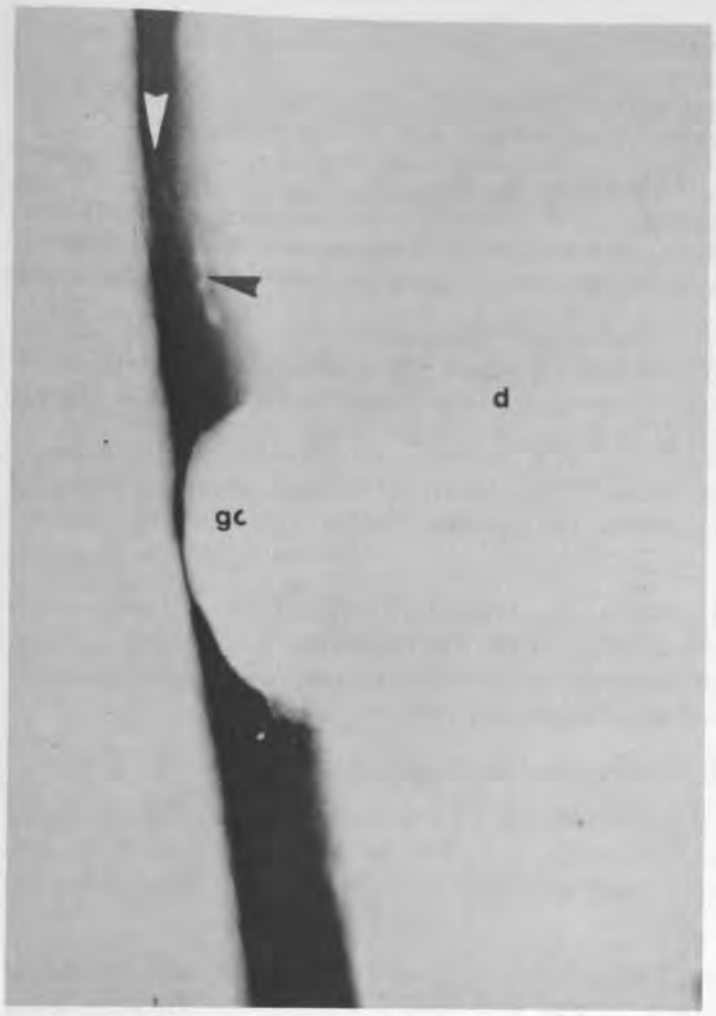

Fig. 6: d=dentin; gc=globular calcification; arrows=needle-shaped calcifications; scale/1 cm=50 μm.
Fig. 6: d=dentine; gc=calcification globulaire; flèches=calcifications en aiguille; échelle/1 cm=50 μm.

Fig. 7: d=dentin; b=bone; arrows=needle-shaped calcifications; scale/1 cm=100 μm.
Fig. 7: d=dentine; b=os; flèches=calcifications en aiguille; échelle/1 cm=100 μm.

Fig. 8: o=osteoplast; nc=nutritional canal; scale/1 cm=15 μm.
Fig. 8: o=ostéoplaste; nc=canal nutritif; échelle/1 cm=15 μm.

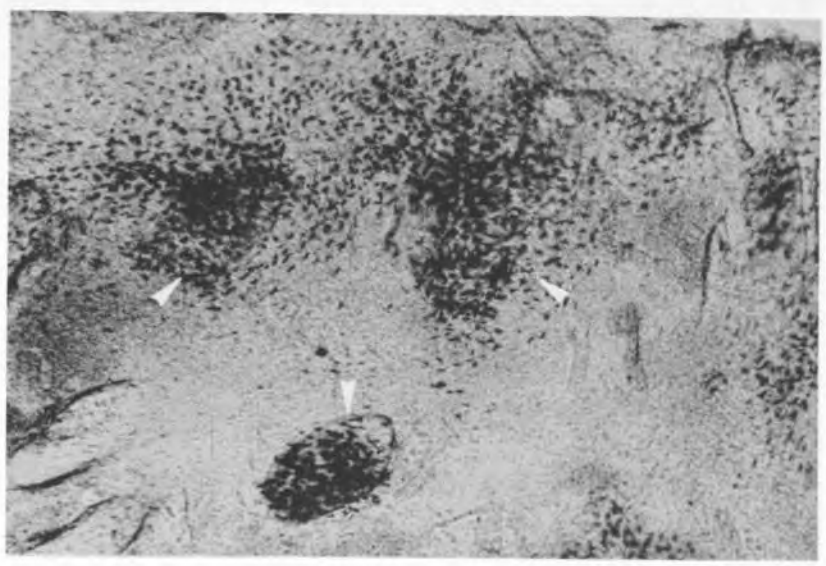

Fig. 9: arrows=three areas of new bone are sited upon resorption lacunae; scale/1 cm=50 μm.
Fig. 9: flèches=trois zones d'os néoformé apparaissent superposées à des lacunes de résorption; échelle/1 cm=50 μm.

cally nine teeth from five cases of cleidocranial dysplasia and showed extreme deficiency of cellular cementum in eight of them. One of these eight teeth was fully and one partly erupted, the others buried.

Histological examination of the roots of primary and permanent teeth showed lack of cellular and acellular cementum (Chapnick and Main, 1976). Little acellular and rarely cellular cementum are present on the roots of permanent teeth in cleidocranial dysplasia but, if they are enable to erupt by surgical exposure, normal acellular and cellular cementum are formed. The acellular cementum of erupted deciduous teeth is, however, normal in thickness and well formed (Hitchin, 1975).

In our case report, distribution of cementum on deciduous teeth would be normal, with presence of acellular cementum on roots and cellular cementum under inter-radicular space and on apex, except that, resorption lacunae seem systematically filled by cellular cementum, so deciduous teeth could remain bound to the alveolar bone.

In patients suffering of cleidocranial dysplasia, Smith (1968) demonstrated that acellular cementum predominated in all of deciduous teeth, with only isolated areas of cellular cementum occuring. Inagreement with previous authors, our study gives prominence to the part play by cementum in eruption and shedding process. According to Hitchin (1975), cementum abnormalities are consistent with a disturbance of bone resorption; we prefer to speak of a disturbance in calcified tissues (bone, cementum) resorption-apposition process.

We are the first author to report intra-pulpal and desmodontal localisations of micro-calcifications in a case of cleidocranial dysplasia. Presence of intra-pulpal calcifications is not surprising in the dental pulp of deciduous teeth (Dard et al., 1988; Arys et al., 1989a; Arys et al., 1989b), but presence of desmodontal mineralization islets is interesting to note and this necessites future investigations. In our opinion, this means a stimulation of mineralisation function, as in bone and cementum, during the disease, probably by defects in genetic regulation process.

This general disturbance of calcified tissues resorption-apposition process may explain the persistance of deciduous teeth and the lack of eruption of permanent teeth in this case of cleidocranial dysplasia.

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REFERENCES

Alderson, C.G.P. — Hereditary cleido-cranial dysostosis. A case report. *British Dental Journal, 108:* 157-159; 1960.

Arys, A., Philippart, C., Dourov, N. — Ultrastructure des microcalcifications du mésenchyme pulpaire des dents temporaires humaines. *Bulletin du Groupement International pour la Recherche Scientifique en Stomatologie et Odontologie, 32:* 23-29; 1989.

Arys, A., Jedwab, J., Pireaux, J.J., Philippart, C., Dourov, N. — Brushite in the pulp of primary molars. *Journal of Oral Pathology and Medicine, 18:* 371-376; 1989.

Chapnick, L.A., Main, J.H.P. — Cementum in cleidocranial dysostosis. *Journal of Canadian Dental Association, 42:* 139-142; 1976.

Dard, M., Kerebel, B., Orly, I., Kerebel, L.M. — Transmission electron microscopy of the morphological relationship between fibroblasts and pulp calcifications in temporary teeth. *Journal of Oral Pathology, 17:* 124-128; 1988.

Eisen, D. Cleidocranial dysostosis. *Radiology, 61:* 21-31; 1953.

Gorlin, R.J., Pindborg, J.J., Cohen, M.M. — Cleidocranial dysplasia. In: Gorlin, R.J., Pindborg, J.J., Cohen, M.M. *Syndromes of the head and neck.* 1975, New York: MacGraw Hill, 180-184.

Hall, R.K., Hyland, A.L. — Combined surgical and orthodontic management of the oral abnormalities in children with cleidocranial dysplasia. *International Journal of Oral Surgery, 7:* 267-273; 1978.

Hasler, J.F., Vandermer, J. — Cleidocranial dysplasia. *Birth Defects, 10:* 524-526; 1974.

Hitchin, A.D., Fairley, J.M. — Dental management in cleidocranial dysostosis. *British Journal of Oral Surgery, 12:* 46-55; 1974.

Hitchin, A.D. — Cementum and other root abnormalities of permanent teeth in cleidocranial dysostosis. *British Dental Journal, 139:* 313-318; 1975.

Järvinen, S. — Dental findings in three cases of cleidocranial dysostosis. *Proceedings of the Finnish Dental Society, 76:* 56-61; 1980.

Jensen, B.L., Kreiborg, S. — Development of the dentition in cleidocranial dysplasia. *Journal of Oral Pathology and Medicine, 19:* 89-93; 1990.

Kalliala, E., Taskinen, P.J. — Cleidocranial dysostosis. Report of six typical cases and one atypical case. *Oral Surgery, Oral Medicine, Oral Pathology, 15:* 808-822; 1962.

Kirson, L.E., Scheiber, R.E., Tomaro, A.J. — Multiple impacted teeth in cleidocranial dysostosis. *Oral Surgery, Oral Medicine, Oral Pathology, 54:* 604-605; 1982.

Magnus, W.W., Sands, N.R. — Cleidocranial dysostosis. Report of a case. *American Journal of Orthodontics, 65:* 638-643; 1974.

Marie, P., Sainton, P. — La dysostose cléido-crânienne héréditaire. *Bulletin de la Société de Médecine des Hôpitaux de Paris,* 436; 1890.

Migliorisi, J.A., Blenkinsopp, P.T. — Oral surgical management of cleidocranial dysostosis. *British Journal of Oral Surgery, 18:* 212-220; 1980.

Monasky, G.E., Winkler, S., Icenhower, J.B., Ruane, A.S., Fielding, A.F., Defrancisis, D. — Cleidocranial dysostosis. Two case reports. *New York State Dental Journal, 49:* 236-238; 1983.

Rushton, M.A. — The failure of eruption in cleido-cranial dysostosis. *British Dental Journal, 63:* 641-645; 1937.

Rushton, M.A. — An anomaly of cementum in cleido-cranial dysostosis. *British Dental Journal, 100:* 81-83; 1956.

Smith, N.H.H. — A histologic study of cementum in a case of cleido-cranial dysostosis. *Oral Surgery, Oral Medicine, Oral Pathology, 25:* 470-478; 1968.

Yamamoto, H., Sakae, T., Davies, J.E. — Cleidocranial dysplasia: a light microscope, electron microscope, and crystallographic study. *Oral Surgery, Oral Medicine, Oral Pathology, 68:* 195-200; 1989.

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