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Chair and Department of Paedodontics Dentistry, Medical University of Lublin

MARIA MIELNIK-BŁASZCZAK, AGNIESZKA ROGOWSKA,
PIOTR STACHURSKI, AGNIESZKA SKAWIŃSKA

Familial incidence of dentinogenesis imperfecta – a case study

Dentinogenesis imperfecta (incomplete development of dentine, Capdepont's illness, opalising dentine), is a genetic disorder inherited in autosomal, dominant way, from one patient to another. There occur asymptomatic carriers of the gene (2, 3). This is the illness developing with the excess of abnormality of mesoderm tissue and is concerned with dentine. It occurs rarely – one case in seven in 10.000 individuals (according to Magnusson) (3). It could be an independently existing illness type II and III (according to Shield) or it could occur as one of the symptoms of osteogenesis *imperfecta* (type I according to Shield) (1, 3, 4, 8). It is concerned with milk and permanent teeth. Clinically the teeth are discoloured into amber-like colour, grayish-yellow-brown and possess characteristic opalisation. The crowns take bell-like or bulging shape, the roots are shortened and disfigured. There occurs partial or total obliteration of tooth cavities, short and narrow canals. The teeth, shortly after coming out, undergo pathological abrasion (3–5). Histological structure in *dentinogenesis imperfecta* is improper. The enamel layer is thin, but composed properly. The dentine in the external part shows proper structure and in the para-ventricular part is improper with the canals of a disturbed shape and course; it has got irregular inter-spherical spaces. One may notice the remaining of the pulp tissues. In the pulp, there occur thick collagen fibres, but odontoblasts are regular. The reason for teeth abrasion is the improper structure of dentine and enamel-dentine junction (1, 3, 6–8).

The aim of the study was the presentation of a familial case of *dentinogenesis imperfecta*. The diagnosis was formulated basing on the detailed family interview and clinical examination.

CASE DESCRIPTION

The patient C. M., aged 13 reported to the Chair and the Department of Developmental Age Dentistry in Lublin in January 2007 with the aim of treatment and prophylaxis of her teeth. The girl presented the proper body structure, the height of 162, but her body mass was low – 37 kg. The girl was born from the first single pregnancy. The girl's development was proper.

From the interview we learn that in the family there have occurred cases of opalising teeth in mother and in other family members – her siblings, grandmother and cousins, grand-grand mother and her siblings, as well as in grand-grandmother's mother (mother's family). The teeth appearance was similar to the one which the girl presented. These individuals all experienced early, developing pathological tooth abrasion (Fig. 1). Extra-oral examination of CM patient did not confirm any problems. The intra-oral examination revealed: erupted permanent teeth, with the exception of third molars. In teeth 14, 26, 36, 37, 45, 46, 47 there are the fillings done. The teeth have characteristic brown-grayish opalising colour, bulging crowns and are shortened as a result of abrasion. It is

especially true about the incisors and first upper and lower molars (Fig. 2a, 2b, 2c). There occurs sensitivity to hot and cold impulses. Pantomographic examination points to the presence in the oral cavity of all permanent teeth with the exception of third molars – their initiations are visible in the bone. The crowns have a bulging shape. There occurs pathological abrasion of the crowns of incisors and of the first molars in both jaws. There is a visible shortening and deformation of the roots. There exists a complete or partial obliteration of cavities and root canals. There are no periapical lesions (Fig. 3). The mother of the child is 37. In her milk and permanent there occurred the symptoms of *dentinogenesis imperfecta*. With age advancement, a gradual loss of hard tooth tissues occurred in the mother; as a result the crowns of teeth underwent abrasion, shortening and became dark-brown. At the age of 26, the child's mother extracted all the teeth in her jaws. The patient C. B. at present possesses an upper, complete, settled prosthesis.

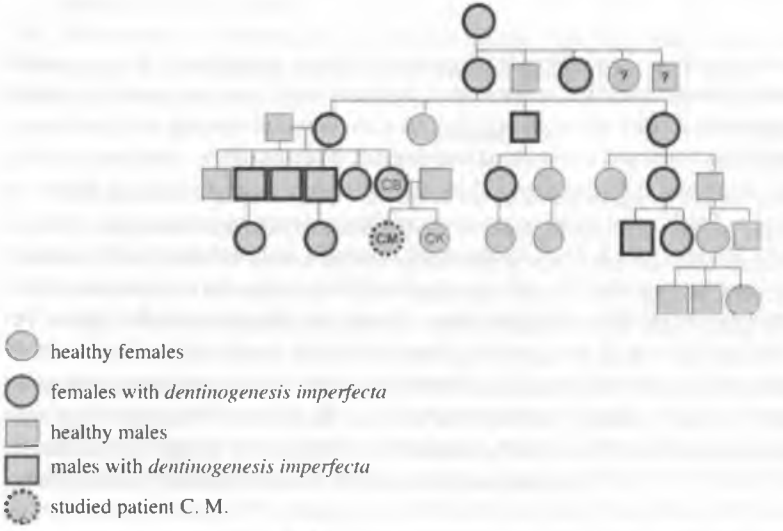

Fig. 1. The scheme of a genealogic incidence of *dentinogenesis imperfecta* in the family of the patient C. M. in five generations done on the basis of the family interview

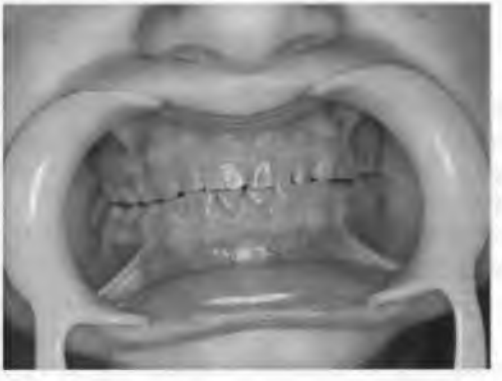

Fig. 2a. Permanent dentition in the 13-year-old patient C. M. – characteristic tooth opalisation of dentine

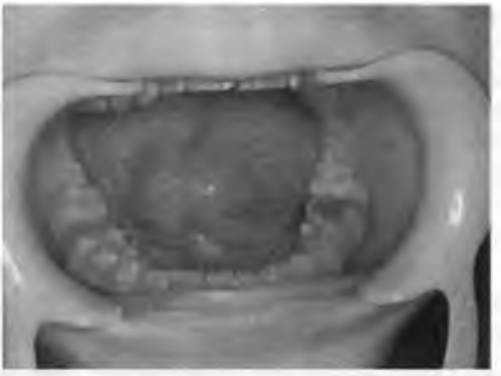

Fig. 2b. The patient C. M. – typical abrasion of the crowns of first molar teeth and incisors in the lower tooth arch

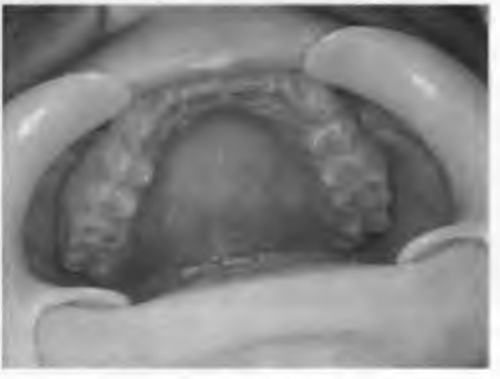

Fig. 2c. The patient C. M. – typical abrasion of the crowns of first molar teeth and incisors in the upper tooth arch

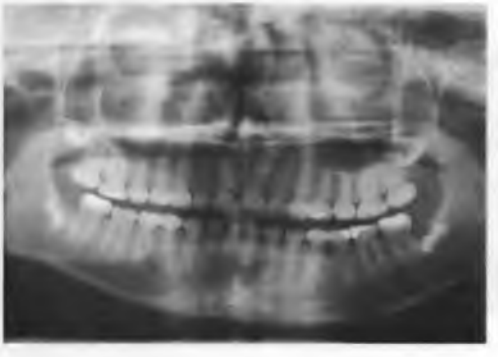

Fig. 3. Pantomographic examination of dentition in the 13-year-old patient C. M. with *dentinogenesis imperfecta*

In the photograph number 4, one may observe a typical *dentinogenesis imperfecta* appearance of her teeth in the lower jaw and the prosthetically elaborated upper jaw (Fig. 4). The patient C. M. has a 12-year-old sister C. K., who was not reported with *dentinogenesis imperfecta*. C. K. has properly formed teeth of a right colour not showing any *dentinogenesis imperfecta* characteristics (Fig 5).

Treatment plan: frequent control examination, instructing in oral hygiene and intense anti-caries prophylaxis, the procedures minimizing tooth over-sensitivity, systematic conservative treatment, the reconstruction of the abrased teeth with composite materials and in the adult age – prosthetic treatment with porcelain crowns. In the case of an advanced crowns abrasion – the use of full or onlay prostheses. Periodic orthodontic control is recommended.

CONCLUSIONS

Dental care of children with *dentinogenesis imperfecta* requires interdisciplinary conduct regarding the developing course of the illness in the whole period of the evolving of the mastication system and after its completion.

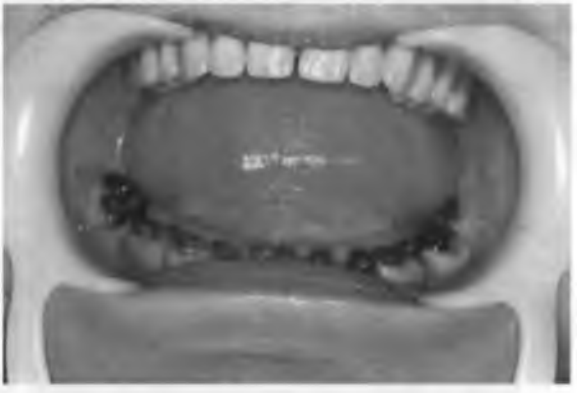

Fig. 4. The appearance of jaw teeth with the lesions typical of *dentinogenesis imperfecta* and prosthetically elaborated jaw in the 37-year-old mother C. B.

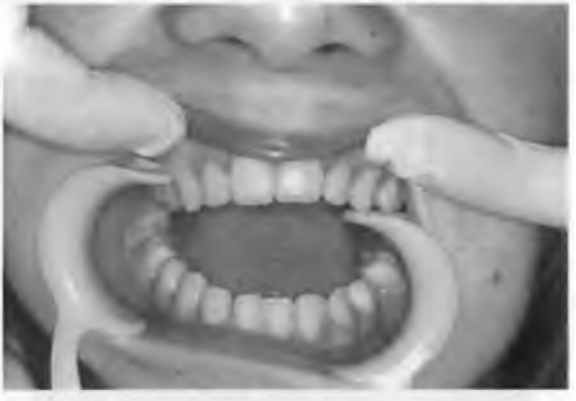

Fig. 5. Proper dentition in the 12-year-old sister C. K. not displaying *dentinogenesis imperfecta* characteristics

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SUMMARY

The aim of the study was the presentation of a familial incidence of *dentinogenesis imperfecta* in five generations. The case of a 13-year-old girl C. M. and of her 37-year-old mother C. B. with the teeth typical of *dentinogenesis imperfecta* was described. *Dentinogenesis imperfecta* is a genetic disease occurring in the family and being inherited dominantly and autosomally. The crowns of milk and permanent teeth have grayish-yellow-brown discoloration and have characteristic opalisation. After coming out they yield to pathological abrasion, the reason for which is the pathological structure of dentine and the enamel-dentine junction. Dental care of children with *dentinogenesis imperfecta* requires multidisciplinary conduct regarding the developing character of the problem in the mastication system evolving and after its full formation.

Rodzinne występowanie *dentinogenesis imperfecta* – opis przypadku

Celem pracy było przedstawienie przypadku rodzinnego występowania *dentinogenesis imperfecta* w pięciu pokoleniach. Opisano przypadek 13-letniej dziewczynki C. M. i jej 37-letniej matki C. B. z uzębieniem typowym dla *dentinogenesis imperfecta*. Jest to zaburzenie genetycznym występujące rodzinnie, dziedziczone autosomalnie dominująco. Korony zębów mlecznych i stałych posiadają szarożółtobrunatne przebarwienie i charakterystyczną opalizację. Wkrótce po wyrznięciu ulegają patologicznemu starciu, którego przyczyną jest wadliwa budowa zębiny i połączenia szkliwno-zębinowego. Opieka stomatologiczna nad dziećmi z *dentinogenesis imperfecta* wymaga postępowania interdyscyplinarnego ze względu na postępujący przebieg schorzenia w całym okresie rozwoju narządu żucia i po jego zakończeniu.