DENTINAL DYSPLASIA

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ONE of the rarest reported dentinal abnormalities is a condition characterized by apparently normal enamel formation with an underlying bizarre, whorl-like dentinal pattern, partial or complete pulpal obliteration, defectively formed roots, and a predisposition to abscess and cyst formation without an obvious inciting factor.

Ballschmiede's[1] description, in 1922, of a case of "rootless teeth" was the first published report of such an entity. Short roots, pulpal occlusion, and early exfoliation were observed in six of seven children in one family. Cysts were found in one of the children and, on gross examination of the exfoliated dentition, apical foramina could be seen. Histologic findings were not reported.

In 1939 Rushton[4] reported the same condition, which he called "dentinal dysplasia," occurring in one person without any indication of familial involvement. Except for peculiar roentgenolucent horizontal lines, the roentgenograms revealed an absence of pulp cavities, defective root formation, and a number of small cysts. Histologic examination showed normal coronal structure and, remarkably enough, the root dentine contained an enormous number of spherical bodies and became more and more irregular toward the apex. In 1954 and 1955 Rushton[5, 6] presented additional material which was illustrated by excellent roentgenographic reproductions and photomicrographs.

In 1951 the occurrence of this same unusual dental condition in nine children and two grandchildren of one family was described by us before the

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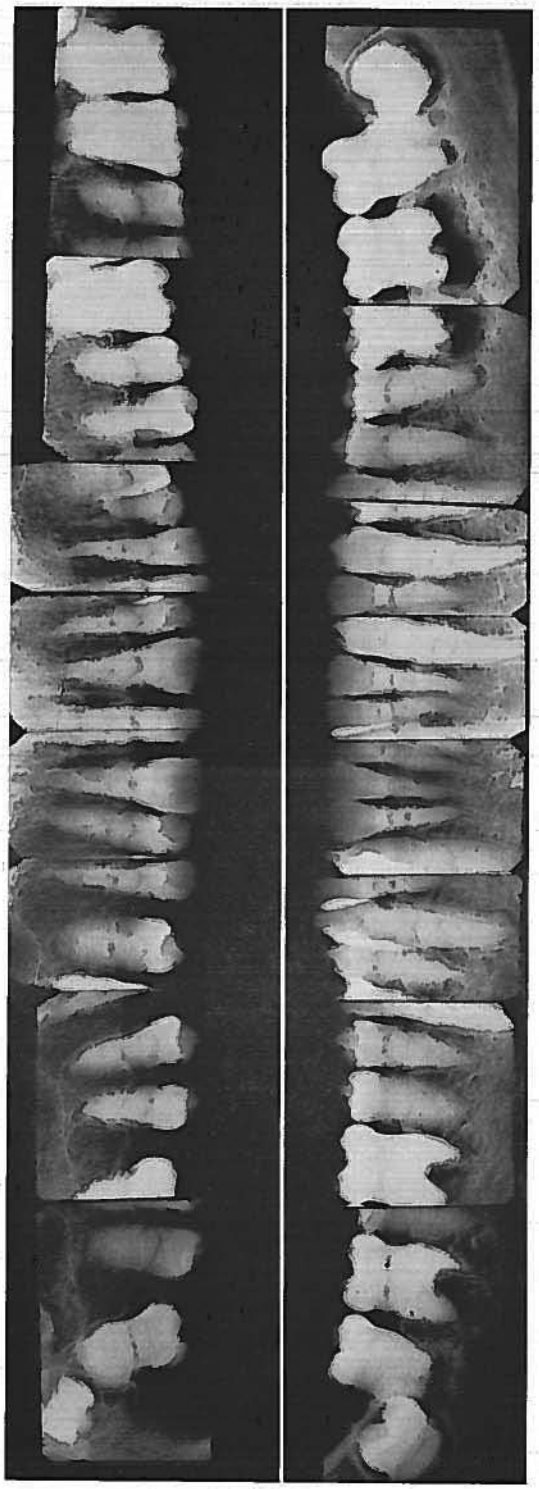


Fig. 1.—Roentgenograms of the 13-year-old son. Occluded pulp chambers with horizontal roentgenolucent lines at various levels are noticeable, with bizarre root formation, areas of apical rarefaction, and somewhat osteoporotic bone. Caries is absent.

Colorado Dental Foundation. However, we delayed publication of our report in order to include the histologic findings in several extracted teeth as well as the roentgenographic and clinical aspects. A detailed analysis is presented here. In the meantime, additional cases have been observed.

In 1952 Hoggins and Marsland[2] reported a case that exhibited features similar to those observed by Rushton—short, bulbous roots with irregular tubular dentine formation interrupted by multiple pulpstones. The pulp tissue was conspicuously absent, enamel formation appeared normal, and the condition was associated with a calcinosis.

In 1957 Zellner[11] reported several families with the same lesions. He examined one family in which three of four daughters were affected while the parents had normal dentitions. The roentgenograms of the three sisters (aged 16, 17, and 18 years) showed the same characteristic features, namely, short roots, obliteration of the pulp, and cystic areas around many apices. The permanent teeth were subjected to careful histologic study with the phase-contrast microscope. The examination revealed the presence of numerous denticles with a varying number of dentinal tubules. The first formed dentine in the coronal portion of the tooth was normal. The roots were covered with a thin layer of cell-free cementum. Zellner found a chronic granulomatous apical periodontitis which he thought was responsible for the apical roentgenolucent areas. He considered the lesion to be a mesodermal formation syndrome and suggested the term "dentinogenesis imperfecta radicularis hereditaria."

Wegner,[8] in 1957, and Wegner and Mannkopf,[10] in 1958, observed two families in which the dentitions showed short roots, pulp obliteration, multiple cyst formation, and early exfoliation. In the first family there were three affected children and their mother, who had an early exfoliation of teeth. The youngest child, who was 9 years of age, showed loosening of the erupted permanent teeth. Also, the deciduous teeth exhibited the typical changes and it was noted roentgenographically that even the unerupted permanent teeth had completely obliterated pulp cavities. In the second family the anomaly was seen in the mother and two children, who had simultaneously a slightly increased serum alkaline phosphatase activity. In the youngest child, a 4-year-old boy, eruption of the deciduous teeth had been somewhat delayed. Additional histologic examination of teeth from this family revealed the same findings as those described by Zellner. In one of the teeth examined histologically, however, Wegner[9] found that the sheath of Hertwig was normal.

The following report, which was presented by us in 1951 and mentioned earlier in this article, concerns nine children of one family and two of four grandchildren who had this anomaly.* Their distinct dental condition was first recognized when one of the sons (13 years of age) reported to the dental office of one of us (J. L.) because of pain in the right maxillary molar area. Clinically, the dentition appeared normal; however, intraoral roentgenograms (Fig. 1) revealed malformed roots, obliterated root canals and pulp chambers

*Several more grandchildren have been born since this first observation was made. Additional records, including the hereditary pattern, may be added at a later date.

Fig. 2.

Fig. 3.

Fig. 2.—Roentgenograms of the 10-year-old son showing evidence of caries along with other features of this type of dentinal dysplasia. Note the peculiar root formation of some of the unerupted teeth.

Fig. 3.—Roentgenograms of the 11-year-old daughter. Note almost complete absence of roots, obliterated pulp structures, multiple apical roentgenolucencies, and poor bone formation. A ground section of the second right mandibular premolar is seen in Fig. 5.

of all teeth, and multiple roentgenolucent periapical areas in the absence of dental caries. Pulp tests seemed to indicate that those teeth with periapical involvement were pulpless. The bone structure was of a porotic nature. Because of the peculiar aspect of the roots and the many roentgenolucencies, which did not resemble those of typical radicular cysts, it was suspected that this condition was an expression of hyperparathyroidism which resulted, even in this abnormal fashion, in an "osteitis fibrosa cystica." The boy was hospitalized for complete medical examination and study, but all metabolic, roentgenographic, and fluoroscopic studies were completely negative.

These findings stimulated an investigation of the dental conditions of other members of the family, especially since some of the other children also complained of "toothache," although in several instances their mouths were caries-free. At the time the first roentgenograms were taken, these children were 20, 18, 15, 13, 11, 10, 8, 6, and 4 years of age. There were six boys and three girls. The four grandchildren (two boys and two girls) were 8, 3, 3, and 2 years of age; only the two boys showed the anomaly.

The father was born in Pennsylvania and the mother in Michigan. The father required very little dental treatment, but the mother had lost her teeth at an early age and wore dentures. She died shortly before this condition came to our attention. The teeth of each of the six boys and three girls were affected, even though clinically they did not appear "unusual."

The roentgenograms of the nine children and two grandchildren revealed a prevalence of teeth with short, bulbous roots that had no more than the faintest traces of a pulp cavity. Only seven full-mouth roentgenograms (Figs. 1, 2, 3, 13, 14, 15, and 16) are shown here as being most characteristic for this distinct type of "dentinal dysplasia," as the remaining four would be repetitious. Most teeth showed horizontal roentgenolucent lines, usually near the base of the crown. There were many apical roentgenolucent areas without evidence of caries or deep restorations. Early exfoliation was characteristic. Erratic response to pulp testing was elicited in some of the teeth that were not involved periapically. The condition in two of the four grandchildren was similar to that found in their aunts and uncles. The family reported no unusual illnesses, and no contributory factors could be detected. There was no apparent indication of a disturbance in the normal pattern of growth and development.

Since the roentgenograms and also the ground sections of several extracted teeth showed no uniformity, a comparison of the individual cases shall be attempted. Roentgenographically, the denture of the 10-year-old boy (Fig. 2) showed this remarkable anomaly already developed before the eruption of the cuspids and premolars. For the most part, the roots of the erupted permanent teeth were short, and some of the mandibular incisors with short roots and large crowns had started already to become loose. This latter phenomenon was most pronounced in the 14-year-old daughter (Fig. 3). The roots were almost completely absent, while the crowns appeared to be of normal size. (Of course, in comparison to the roots, the crowns appeared

to be large.) Most of the teeth were loose and were subsequently extracted. Ground sections of a mandibular molar and premolar from Fig. 3 are shown in Figs. 4 and 5.

In general, it can be said that histologically (and also clinically) the enamel appears normal, with some thinning seen in cervical areas. As noted in the ground sections shown in Figs. 4 to 7, the dentinal tubular pattern in the coronal portion appears to start out in a normal, organized manner, even though this primary dentine layer under the enamel appears irregular in several areas and sometimes atubular and one is reminded of the dentine formation in dentinogenesis imperfecta. The granular layer of Tomes can be recognized clearly in Figs. 5 and 6. Centrally and apically, this pattern is interrupted by irregular, bizarre, whorl-like structures suggesting multiple pulpstones (Figs. 5 and 6), which completely obliterate any semblance of pulp chamber or canals. The horizontal lines, which are clearly visible in the roentgenograms, can be recognized in Figs. 4, 5, 6, and 7; these seem to represent the areas which were originally filled with pulp tissue and which, of course, were lost during the preparation of the ground sections. Nevertheless, as can be seen in Fig. 4, there are several of these horizontal spaces, and one gains the impression that they are incremental lines of wild-growing dentine formation.

The histogenetic pattern of dentine formation is by no means always the same, as demonstrated in Figs. 6 and 7. In most instances the cementum is extremely thin, and the short, pointed root suggests the failure of Hertwig's sheath to develop a normal length.

A mesiodistal ground section of a deciduous second molar (Fig. 8) shows a fine pattern similar to that seen in Fig. 7. In this particular case the irregular whorl formation has already begun in the pulpal horns. Areas *a*, *b*, and *c* in Fig. 8 are shown at a higher magnification in Figs. 9, 10, and 11. Here it should be noted that the dentine in certain areas is quite atubular. The "whorls" seem to form particularly in those areas (*a* and *b* in Fig. 8) where the dentinal tubules are numerous (Figs. 9 and 10). Fig. 11 (*c* in Fig. 8) again shows this peculiar grotesque and irregular dentine formation which does not resemble pulpstones and seems to have grown completely out of hand. A similar distinctive pattern may be seen in Fig. 12, which shows a central mesiodistal ground section of a deciduous molar.

Roentgenograms of some of the other children in the family (Figs. 13, 14, and 15) show pointed roots similar to those seen in Fig. 3, with multiple periapical roentgenolucencies. Distinct horizontal roentgenolucent lines which actually show pulpstones may be seen in Fig. 14.

The roentgenograms of the 4-year-old grandson (Fig. 16) show that the anomaly affects deciduous as well as permanent teeth and that the unerupted permanent teeth show the involvement already present long before their eruption. The fact that the child is a grandson emphasizes the hereditary aspect of this condition.

Cross sections through the coronal and cervical portions of a cuspid are

Fig. 4.

Fig. 5.

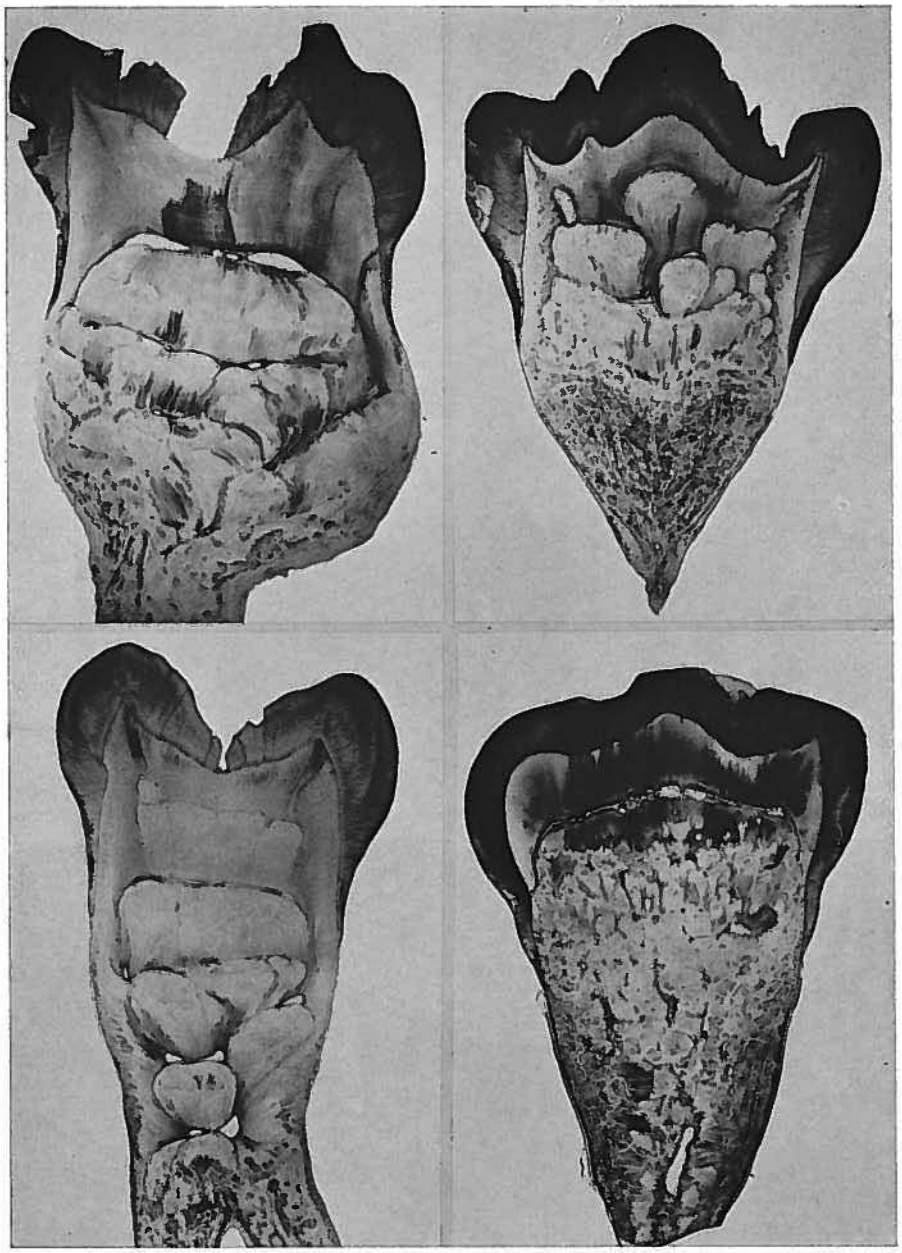


Fig. 6.

Fig. 7.

Fig. 4.—Ground section of a permanent upper second molar shows the typical bulged-out, stubby, root appearance. Normal-appearing enamel, dentine, and cementum are observed peripheral to the central bizarre configurations filling the entire pulp cavity.

Fig. 5.—Ground section of the right mandibular second premolar of roentgenogram shown in Fig. 3. Central dentine formation resembles that of pulpstones.

Fig. 6.—Ground section of a slender upper right first premolar with some semblance of bifurcation. Peripheral dentine is of normal structure. Central pulpal obliteration with dentinal whorls (pulpstones) becomes completely disorganized apically. Spaces seen probably represent loss of organic material during sectioning.

Fig. 7.—Ground section of the right mandibular molar of roentgenogram shown in Fig. 1. Note the different and smaller pattern of the dysplasia in contrast to Figs. 4, 5, and 6.

Fig. 8.

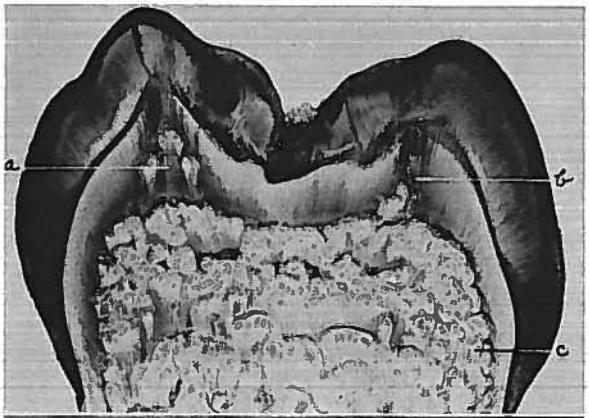
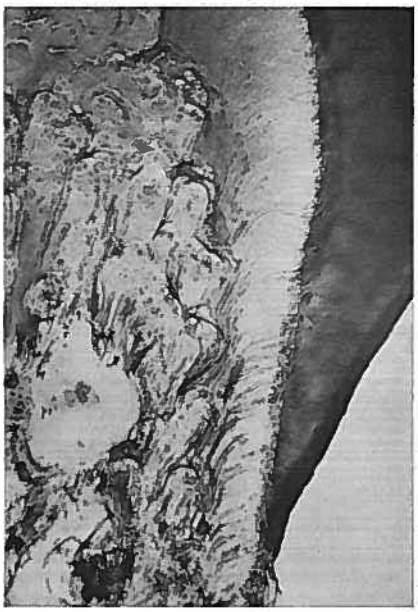


Fig. 9.

Fig. 10.

Fig. 11.



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seen in Figs. 17 and 18; it is evident here that vessel spaces are actually present, even though they are extremely small. The section shown in Fig. 17 was made near the cementoenamel junction, and the one shown in Fig. 18 was made through the pulp chamber. A section through the root portion of another tooth (Fig. 19) shows some primary dentine and a completely disorganized deposition of dentinoglobules which obliterate the entire pulp canal.

Again, in these ground sections there can be seen several empty spaces, which were examined more closely in histologic sections of several teeth. They showed uniformly connective-tissue elements and blood vessels. For the most part, the capillaries were empty (Figs. 20 and 21).

Only a few normal odontoblasts could be found, while mostly degenerative changes resembling those of a "reticular atrophy" prevailed. The possibility of shrinkage of the specimen during the histologic preparation cannot be entirely excluded in the interpretation.

Finally, the roentgenograms of the father's mandibular anterior teeth, from first premolar to first premolar, are shown in Fig. 22, with no sign of the described anomaly.

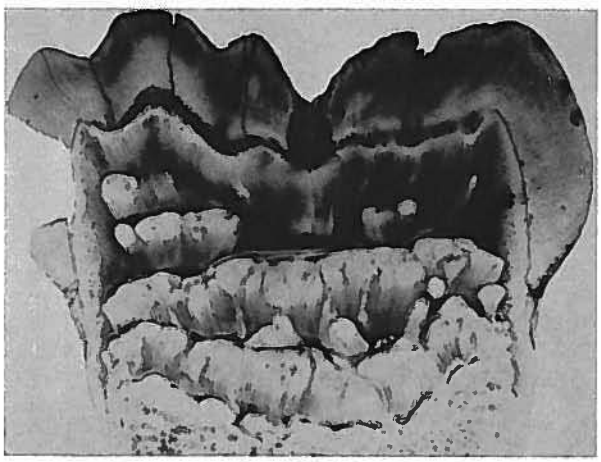

Fig. 12.—Central mesiodistal ground section of a deciduous molar, similar to that seen in Fig. 4.

DISCUSSION

On the basis of the eleven cases reported here and the eighteen cases cited from the literature, it seems possible to give a fairly good description of this very rare anomaly. No sex preponderance seems to exist. The condi-

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Fig. 8.—Ground section, cut in a mesiodistal direction, of a deciduous lower second molar of 4-year-old daughter. Normal enamel formation is evident. Coronal dentine has a normal beginning, rootward becoming a tortuous mass of irregular dentinal whorls, obliterating the pulpal area.

Figs. 9, 10, and 11.—Higher magnifications of areas *a*, *b*, and *c* of Fig. 8. Note the irregular tubular arrangement.

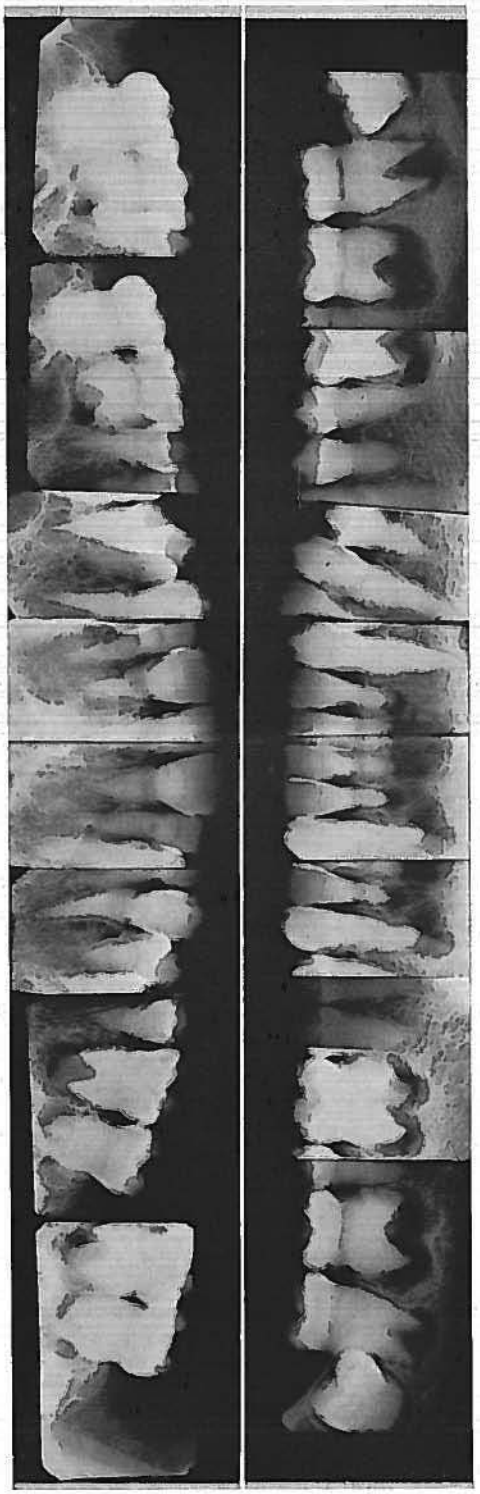

Fig. 13.—Roentgenograms of 18-year-old son showing irregular root formation with multiple periapical roentgenolucencies.

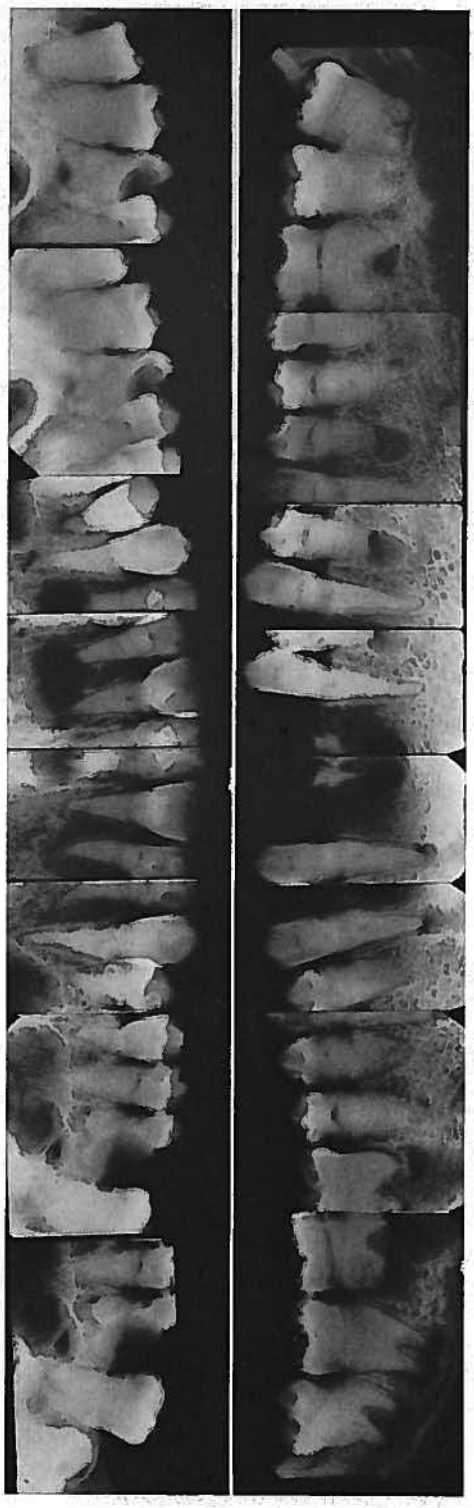

Fig. 14.—Roentgenograms of 20-year-old son showing areas of advanced decay and extreme periapical involvements.

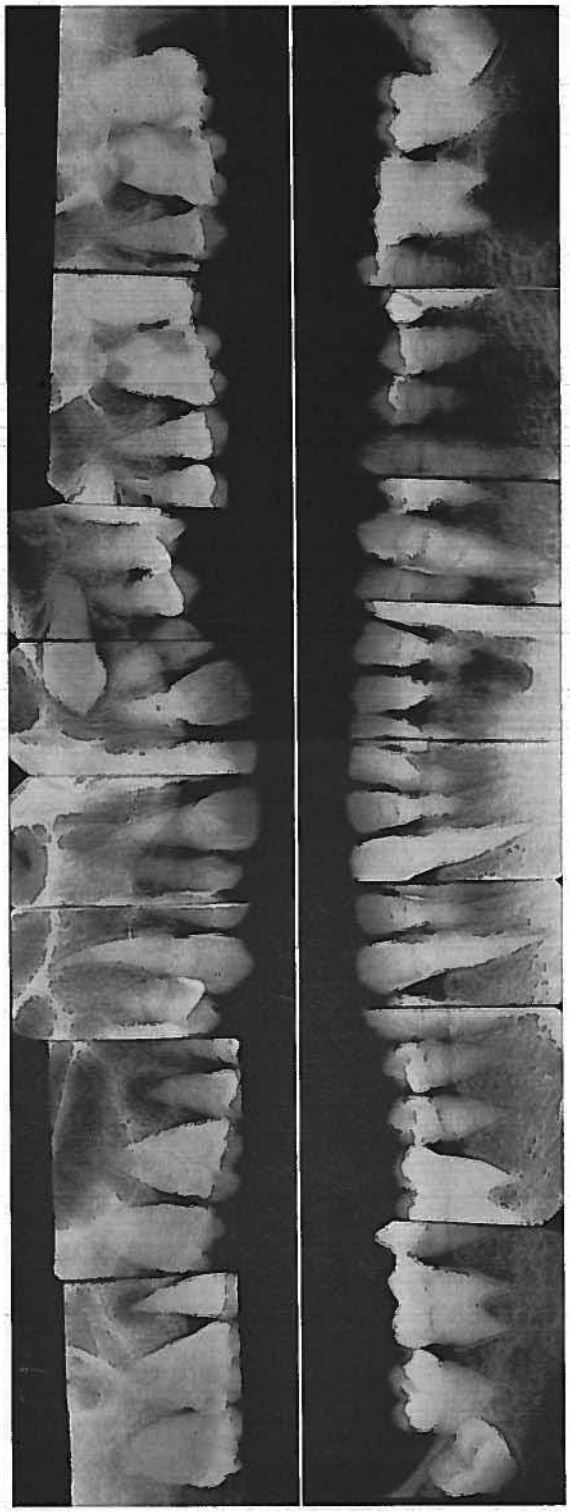

Fig. 15.—Roentgenograms of 15-year-old son showing similar pattern of pointed root formation, obliteration of pulp canal and multiple periapical roentgenolucencies. Bone structure appears normal.

tion is hereditary, a concept which is supported by the occurrence of the lesion in both deciduous and permanent teeth in fathers, mothers, and children.

Clinically, the dysplasia is characterized by loosening of the teeth, which leads to an early exfoliation, apparently because of the relatively large crowns and short, pointed roots, and the development of periapical infections or cysts. The teeth seem to be somewhat caries-resistant. The teeth erupt at normal times, although a few cases of slightly delayed eruption have been reported. No systemic background has yet been demonstrated in connection with the anomaly; nor has there been a deviation from the normal growth

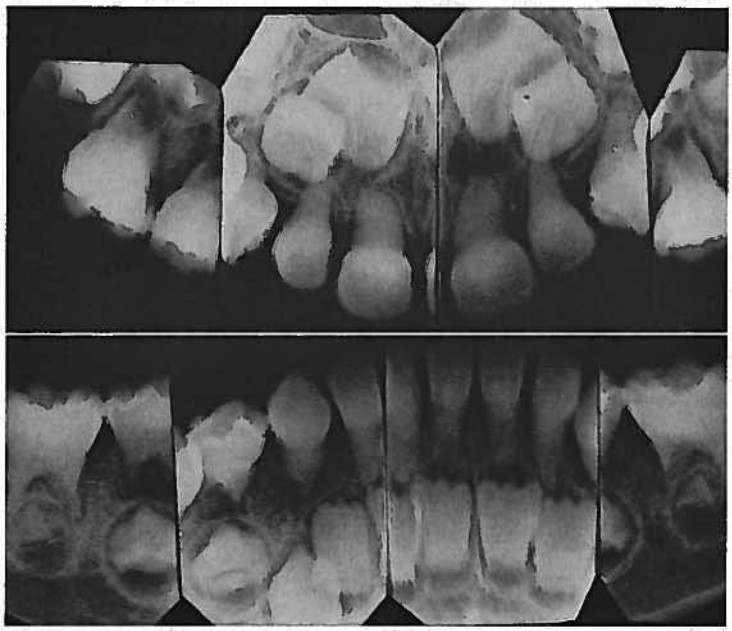

Fig. 16.—Roentgenograms showing the affected deciduous dentition and even the unerupted permanent teeth of one grandson. Malformed roots and obliterated pulp chambers and canals are evident.

or developmental pattern of these children. Clinically, the crowns of the teeth do not show any deviations from the normal with regard to color or to morphology. In some cases the gingivae were found to be inflamed, which appears to be coincidental. Usually the teeth showed a low threshold value on electric pulp testing. Often the patients consulted a dentist only because of "pain," and a roentgenologic examination disclosed the abnormal pulp condition.

Roentgenographically, the affected teeth present a peculiar picture. Most roots are very short, with a strong tendency to periapical involvement. The pulp cavities are obliterated, and only scattered remnants of pulp tissue can be observed. Most teeth show horizontal, narrow, roentgenolucent lines, usually near the base of the crown. It should be emphasized that the pulp cavities are obliterated even before the teeth erupt into the oral cavity.

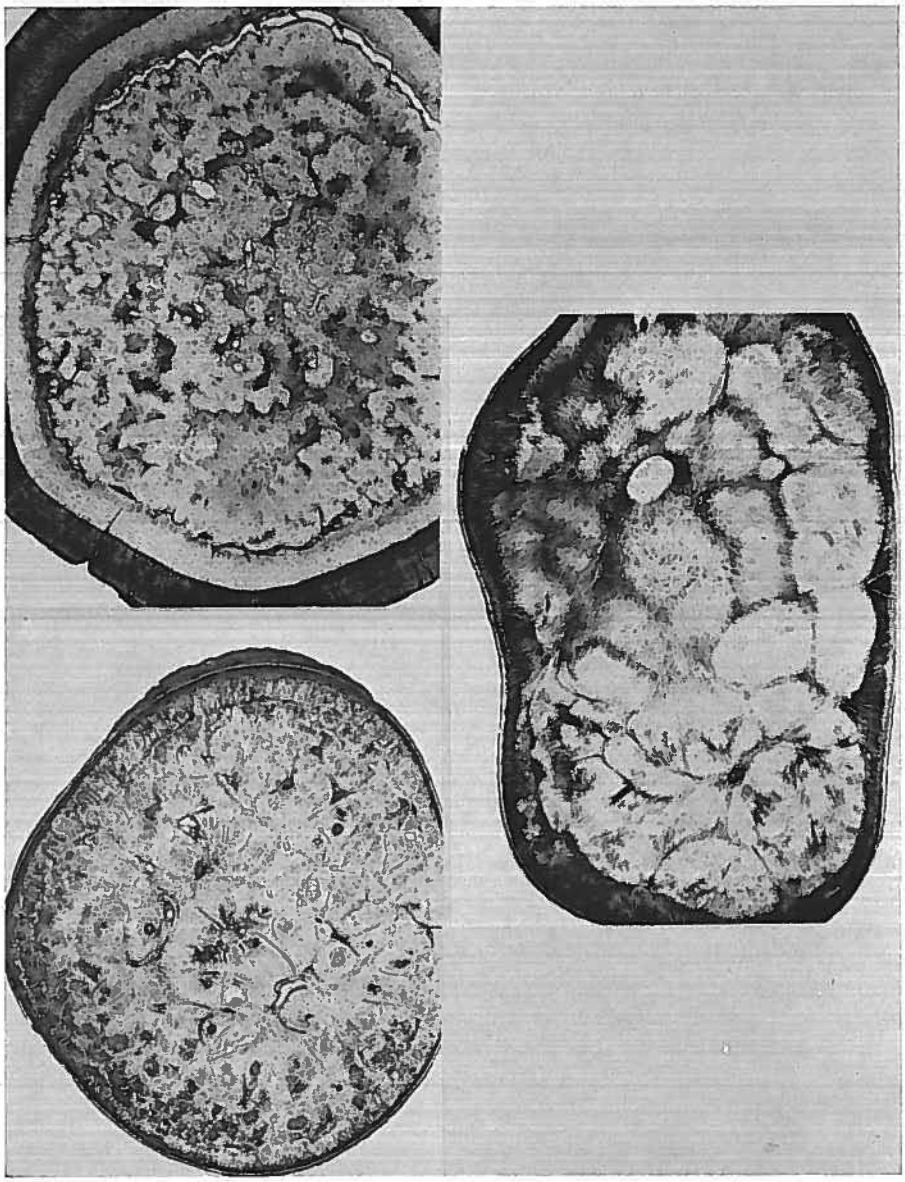

Fig. 17.

Fig. 18.

Fig. 19.

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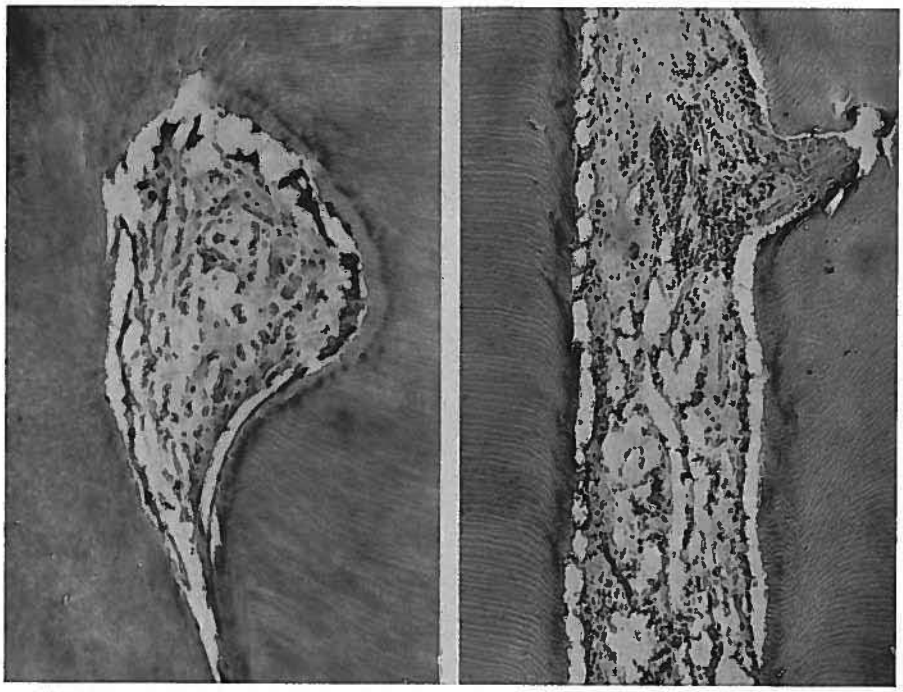

Fig. 20. Fig. 21.

Figs. 20 and 21.—Photomicrographs of remnants of pulp tissue showing partially empty capillaries and degenerative changes of the odontoblastic layer.

Histologically, the teeth reveal a varying but typical and unique structure. The first formed dentine in the coronal part appears normal, even though in most instances the tubular arrangement is irregular. The rest of the dentine and the pulp cavity are filled with irregular, bizarre, whorl-like

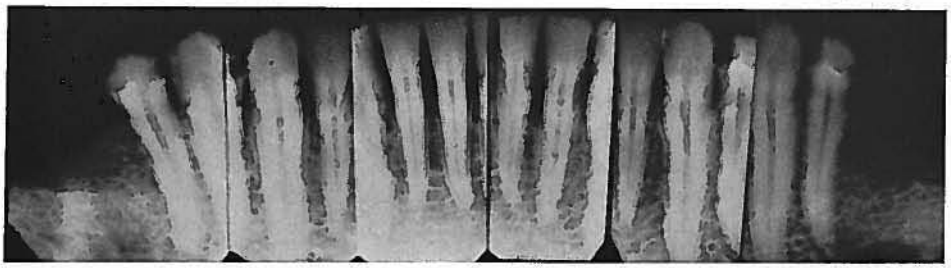

Fig. 22.—Normal dentition of father.

Fig. 17.—Cross section at cervical line of deciduous upper cuspid of 6-year-old son; note presence of some normal enamel and cementum. Central to this is wild, disorganized dentine laid down in whorl-like configurations.

Fig. 18.—Cross section of the same deciduous cuspid as shown in Fig. 17 halfway between the incisal surface and the cementoenamel junction. The enamel appears normal, but the dysplasia pattern is different from that seen in Fig. 17, which was taken from the cementoenamel junction area.

Fig. 19.—Cross section through the root of a permanent molar demonstrating the peculiar "pulpstone-like" pattern of the dysplasia.

structures (denticles) with a varying number of dentinal tubules. These spherical bodies obliterate the pulp almost totally. The disorganization of the hard-tissue structure increases apically. In a few cases the content of the cystic cavity has been examined histologically and found to be lined with ordinary epithelial cystic membranes.

The anomaly is designated as "dentinal dysplasia," a term first used by Rushton[4] in 1939.

In comparing the dentinogenesis imperfecta with the dentinal dysplasia, both of which show drastic alterations in dentinal development, we should point out that the two conditions have certain features in common. Both are hereditary and affect the deciduous as well as the permanent dentitions. In both instances the enamel appears to be normal. There are also striking differences, however; for instance, the teeth in dentinal dysplasia are not opalescent and do not show any degree of the attrition seen in dentinogenesis imperfecta. Both conditions show obliteration of the pulp cavities, but this phenomenon occurs much earlier in dentinal dysplasia than in dentinogenesis imperfecta (in fact, even before tooth eruption). A common feature is the normal deposition of the first layer of dentine in the two lesions, but the pathologic dentine formation that follows is different, with formation of spherical bodies dominating in dentinal dysplasia. However, Schulze[7] has published some photomicrographs from dentinogenesis imperfecta teeth that had some spherical bodies. This might be an indication that transitional cases between the two anomalies may exist. This postulate is supported by the findings in the triracial group in Maryland reported by Hursey and associates.[3] In this group they found a high incidence of dentinogenesis imperfecta and stressed that the disease in the group shows a wide variation in its clinical, roentgenologic, and histologic appearance. Hursey and his colleagues are of the opinion that the process of dentine formation may be arrested at various stages, resulting in a defect called "shell" teeth or in a pulp chamber of intermediate size.

It has been suggested that the deformed roots in dentinal dysplasia are due to an altered activity of Hertwig's sheath in producing a normal structure. However, the only known histologic examination of Hertwig's sheath in a case of dentinal dysplasia (Wegner's case[8]) showed no signs of deviation from the normal. The etiology appears to be related to multiple foci of degeneration in the dental papilla as a principal factor leading to reduced growth and final obliteration of the papilla and to the formation of sporadic true dentine about calcified foci.

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