CONGENITAL MALFORMATIONS IN THE TEETH AND EYES IN MENTAL DEFECTIVES.

By RICHARD SPITZER, D.M.D., and

IDA MANN, D.Sc., M.B., B.S.(Lond.), F.R.C.S.(Eng.).

THE interest in developmental abnormalities and their aetiology has considerably increased in recent years. In spite of this, there is a paucity of information about congenital anomalies of the teeth and about their association with similar lesions in other parts of the body.

The present paper deals with dental malformations as part of a syndrome comprising other congenital lesions, and mental deficiency.

Dental dystrophies are due to various causes acting in the pre- or post-natal period, and the deformities are variable in extent and degree. Most of the clinical and experimental work deals with the effects of post-natal environmental factors on the tooth structure, and the study of the aetiology of the congenital dental lesions has been relatively neglected (Moore, 1944).

The enamel of the tooth, the lens of the eye and the nervous tissue are among others, derivatives of the ectodermal layer. It is well known that various clinical syndromes may arise in one or several of these structures from faulty development. This may be due either to a primary lesion in the embryonic layer or to an interference with the developmental process early in foetal life. An example of the former is hereditary ectodermal dysplasia, a condition characterized by defects in one or several organs arising from the ectoderm. The dental manifestations are malformations of the tooth structure and partial or complete anodontia. The example of the latter aetiology is congenital syphilis, where the lesions of the permanent teeth can be pathognomonic. The enamel deficiency is present before the eruption of the teeth as observed by histological and radiological observations (Sarnat and Schour, 1941; Bauer, 1944). The present investigation was carried out to find whether dental malformations can be related to the pathology of the endogenous mental disease. A further problem studied was the incidence of developmental lens defects in these patients. This was done to see whether such changes would support the argument that the causative agent or agents are partly or wholly synergistic.

The assumption that congenital syndromes can often be reduced to ectodermal, mesodermal and entodermal lesions, has in recent years been questioned. In its place has been suggested a classification based on biochemical (enzymatic) affinities (Grüneberg, 1947). The present work, however, is to some extent based on the first mentioned hypothesis which, whatever its demerits, has not proved unfruitful, since the biochemical affinities probably correspond to a

certain extent with the histological and is still affirmed by many authors (Boyd, 1943; Bruno and Engelhardt, 1944).

In this paper abnormalities of the permanent teeth only are considered. The permanent teeth erupting six or more years after the factors of any maldevelopment have ceased to act, still exhibit the characteristic pattern of the maldevelopment. There are other congential or inherited diseases whose clinical manifestations are only seen years after birth, e.g. retinitis pigmentosa, Friedreich's ataxia, etc. (Grüneberg, 1947).

In varying degrees other congenital systemic diseases, in addition to those previously mentioned, may provoke structural anomalies of the dentition.

In the last few years maternal rubella has been identified as a cause of congenital malformations in the child (Gregg, 1941; Swan, Tostevin *et al.*, 1943, 1946), and since then evidence has accumulated to confirm these observations. The effects on the child are most marked in cases where the mother contracted the disease between the sixth to ninth week of pregnancy. Evans (1944, 1947) was the first to describe the developmental defects in the deciduous teeth in this condition. His findings indicate a close aetiological relationship between the dental defects, and the other congenital lesions such as cataract, deaf-mutism, microcephaly, mental deficiency resulting from the systemic infection. They are, furthermore, an attempt to correlate the dental changes with the clinical history and to assess the time of exposure to the injurious agent within narrow limits.

More information has been collected about tooth anomalies associated with congenital lesions of the skin, and some of it has to be mentioned briefly. Cockayne (1933) reviews the whole question and points to various forms of dental maldevelopment in this condition, e.g. enamel hypoplasia, microdontia, partial or complete anodontia and delayed dentition. He also discusses their association with lens changes and mental defects. His views are substantiated by many other authors (Halperin and Curtis, 1942; Bruno and Engelhardt, 1944; Andrews, 1946; and Sequeira, Ingram and Brain, 1947), and from the data submitted there is sufficient evidence to state that the unusual dental features are due to the same early developmental or germinal deficiency which also produces the other defects.

Consequently, we can see that a single pathological aetiology, not only may provoke changes in tooth, skin and lens, but, in addition, mental deficiency.

As far as one of us (R. S.) is aware, no large-scale inquiry has been made into this question. Tredgold (1947) remarks on the high incidence of dental disorders in various groups of mental defectives, especially in the presence of a palatal deformity. Of particular interest are the reports on dental anomalies in mongolism. Tooth lesions in this condition were recognized by Jones (1890), quoted by Brousseau (1928). Kreyenberg (1936), van der Scheer (1927), Thomas (1939) and Benda (1947) have, among others, also noted the frequent occurrence of various forms of dental dystrophies, such as hypoplasia, malformed teeth, microdontia, anodontia and delayed eruption.

This survey has shown that there are instances where the tooth anomalies assume significance as part of the clinical syndrome.

In some of the mentioned systemic diseases the associated dental lesions

are clearly not due to genetic causes, but to foetal damage (congenital syphilis, maternal rubella). If dental anomalies are found in patients affected with genetically inherited diseases, it is necessary to examine whether these tooth defects are also genetically determined or are due to other factors.

The significance of this point is obvious. Snyder (1941) and more recently Gates (1946), have recognized that the dental tissues in their undifferentiated state are subjected to genetical influences. However, since the dental stigmata are not characteristic enough to indicate the aetiology, it may be of help to look for related changes in other parts of the body. The findings can then be used as a guide in clarifying the problem.

Previous investigations of the occurrence of dental lesions in mental patients have not attempted to differentiate these defects into groups, and did not segregate the cases with dental and other developmental anomalies from those without these conditions.

A high incidence of hypoplasia of the tooth enamel in idiopathic epilepsy has previously been reported (Spitzer, 1933, 1942). The present paper deals with a larger number of patients, and the investigation was not confined to cases of idiopathic epilepsy, but included mongols and unclassified mental deficient patients in order to find the incidence of dental defects in such inborn cerebral diseases and to trace the connection with congenital lesions in other organs of ectodermal origin.

There were two important reasons for selecting mental cases for this purpose. Firstly, among such cases there are many whose aetiology is known to be genetical, producing pre-natal lesions in other organs; and secondly, the tooth enamel shares with the nervous system its origin from the ectodermal layer.

The present study aims to clarify the following points:

1. Which dental anomalies can be considered congenital.

2. The incidence of dental lesions among all three groups of mental defectives examined:

   (*a*) Idiopathic epilepsy.
   (*b*) Mongolism.
   (*c*) Unclassified mental deficiency.

3. The incidence of dental and lenticular lesions within these three groups.

4. The incidence of associated dental and lenticular lesions within each of these groups.

Clinical Findings.

Two-hundred and thirty patients in Leavesden Hospital and 89 patients in the Fountain Hospital were examined. The examination was augmented by checking the history of each patient, including his family history, in order to trace genetic and acquired influences.

Eighty-three of the patients in Leavesden Hospital had their eyes examined by Professor Ida Mann. They were divided into two groups, one with dental lesions, and the other without, but both having the same mental disease.

Only those cases showing bilateral lesions of the teeth were considered positive. Where the contralateral tooth was missing or did not permit of a

definite diagnosis, or where the condition was not sufficiently marked, the case was classified as doubtful.

Three different types of dental disorders were noted:

1. Enamel hypoplasia.

2. Malformation of shape, the outstanding features of which are either the shortening of the transverse axis, producing long and narrow teeth, or the loss of the normal incisal edge, producing peg-shaped teeth.

3. Microdontia.

All patients were excluded whose mental condition was obviously the result of birth injuries, trauma in early infancy or congenital syphilis.

Among the 83 cases who were examined dentally and ophthalmologically, the ages of the imbeciles ranged from 24 to 47, of the mongols from 21 to 43, and of the idiopathic epileptics from 19 to 64.

Enamel hypoplasia was present in all three types, though the highest incidence was in idiopathic epileptics and imbeciles. Malformed, peg-shaped teeth and microdontia were most prevalent in mongols.

The 230 cases in Leavesden Hospital (Tables I, II, III) comprised 100 idiopathic epileptics, 105 unclassified mental deficient patients, among which were 2 cretins, and 25 mongols.

Among the 89 cases in the Fountain Hospital (Tables IV, V, VI) 15 were idiopathic epileptics, 61 were unclassified mental deficient patients, one of whom was a patient with congenital syphilis, and 13 were mongols.

Of the 230 cases in Leavesden Hospital, 64 showed dental malformations of various types, 13 were doubtful, and 153 had normal teeth. Of the 89 cases in Fountain Hospital 20 patients exhibited dental lesions, 2 patients had to be regarded as doubtful, and 67 had normal teeth.

The 83 cases who were examined ophthalmologically as well as dentally were grouped according to their psychiatric diagnosis, and then subdivided according to the positive or negative findings in their teeth. In these groups there were:

51 idiopathic epileptics (Tables VIIA, B, C).

19 mongols (Tables VIIIA, B).

13 unclassified mental deficient patients (Tables IXA, B).

Among the 51 idiopathic epileptics (Tables VIIA, B, C, and Table X) the results were:

Teeth:		*Eyes:*	
Positive	27 (53%)	Positive	24 (47%)
Doubtful	5 (10%)	(1 due to congenital syphilis).	
Negative	19 (37%)	Negative	27 (53%)

Among the 19 mongols (Tables VIIIA, B and Table XI) the results were:

Teeth:		*Eyes:*	
Positive	16 (84%)	Positive	14 (74%)
Doubtful	1 (5%)	Doubtful	1 (5%)
Negative	2 (11%)	Negative	4 (21%)

https://doi.org/10.1192/bjp.96.404.681 Published online by Cambridge University Press

Among the 13 unclassified mental deficient patients (Tables IXA, B and Table XII) the incidence was as follows:

Teeth:		*Eyes:*	
Positive	9 (70%)	Positive	5 (38%)
		Doubtful	1 (8%)
Negative	4 (30%)	Negative	7 (54%)

Sex was not found to influence the dental condition.

The observations show that the highest incidence of dental and lenticular lesions is found in mongols.

The typical dental changes in mongols were the malformation of shape producing the haplodont or peg-shaped tooth, and the smallness of the permanent dentition (microdontia), giving the appearance of deciduous teeth, and frequent discrepancy in size between the various groups of teeth, particularly between the lateral and central incisors (Figs. 1, 2).

The commonest defect, hypoplasia of the enamel, encountered in the idiopathic epileptic and unclassified mental deficient group is characterized by pitting and grooving of the enamel along the developmental lines. This defect persists at different levels in different groups of teeth, due to the fact that the causes for the anomalies take effect at a certain growth period involving, therefore, various tooth groups at different stages of development. (Figs. 3, 4, 5, 6). Hence, the lesions will be found nearer to the incisal edge or occlusal surface, which is the first part of the tooth to form, in some groups, in others they will be farther away. Of the other defects, only on one occasion was microdontia found in idiopathic epileptics and unclassified mental deficient patients. As far as can be ascertained it appeared that the incidence of dental disorders was more marked in those patients whose family history showed other cases of mental deficiency (Figs. 3, 4, 5, 6). It was frequently found, furthermore, that where there were birth injuries, traumata or post-natal disease, those patients did not show any evidence of dental, or lenticular pathological development.

The eye changes observed in the 83 patients examined with the slit lamp and ophthalmoscope were mostly lenticular, as appears from the tables and statistics given above. A few other congenital anomalies of minor import were also found, namely incomplete persistence of hyaloid artery (2 cases), persistent pupillary membrane (2 cases), opaque nerve fibres, inferior crescent, pigment ring round optic disc, gliosis of disc and choroideraemia (one case each). Acquired eye conditions were few, but posterior synechiae (3 cases), traumatic cataract, dislocation of one lens, trachomatous pannus and secondary optic atrophy (one case each) were found.

The changes in the lenses, apart from the 2 cases above, were all of developmental type. They were most frequent in the group of mongols and included the conditions described and depicted by Lowe (1949), namely *arcuate opacities* due to maldevelopment of the lens fibres themselves, and arising between the second and third month of intra-uterine life, *dot-like or flake opacities* outside the foetal nucleus, of coronary type due to various lacunae and vacuoles

between the lens fibres and arising during post-natal life, and *suture cataracts* due to deposition of opaque material in the *Y* sutures of the foetal nucleus, and therefore ante-natal (third month onwards). The coronary opacities are frequently found in normal individuals, but among mongols their incidence

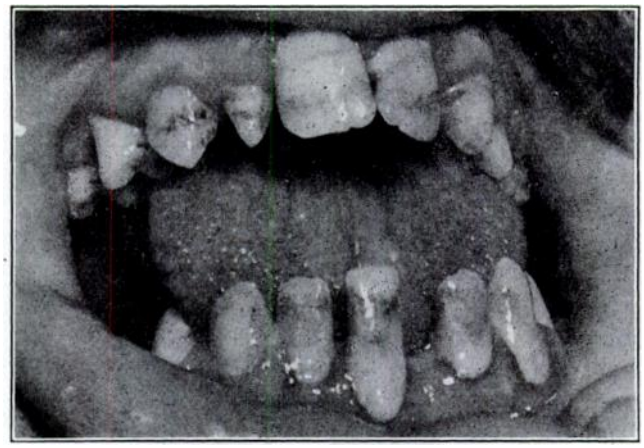

FIG. 1.—H. F—, age 27. Mongol (imbecile). M.A. 4, 6 years, I.Q. 32. Teeth: Malformed and peg-shaped. Eyes: Positive findings. Speech: Defective. Palate: High.

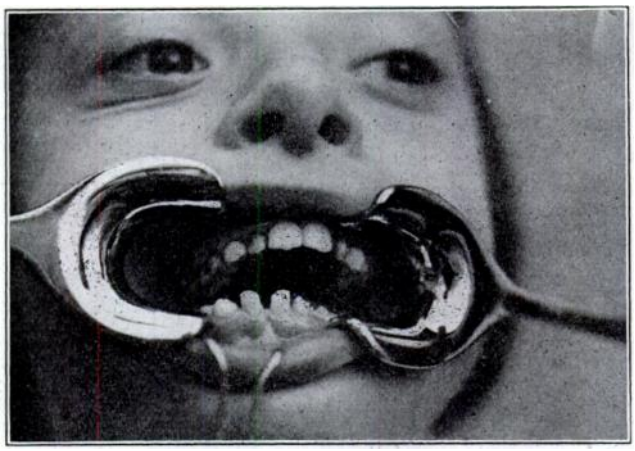

FIG. 2.—D. W—, age 18. Mongol. M.A. 3, 4 years, I.Q. 31. Microdontia. Palate: High. Tongue: Mongolian (fissured). Small last fingers on both hands.

is higher than would have been expected. Even mild cases in the mongols were associated with defective tooth development.

In some of the mongols with coronary opacities there were also brightly-coloured crystalline dots in the cortex of the type usually labelled "endocrine cataract." All this agrees with Lowe's findings, that many different types of

lenticular maldevelopment can be found in mongols, and it is further interesting to note their almost constant association with dental abnormality.

In the other groups (idiopathic epileptic and unclassified mental deficient patient) lens opacities did not occur in so high a proportion, but their associa-

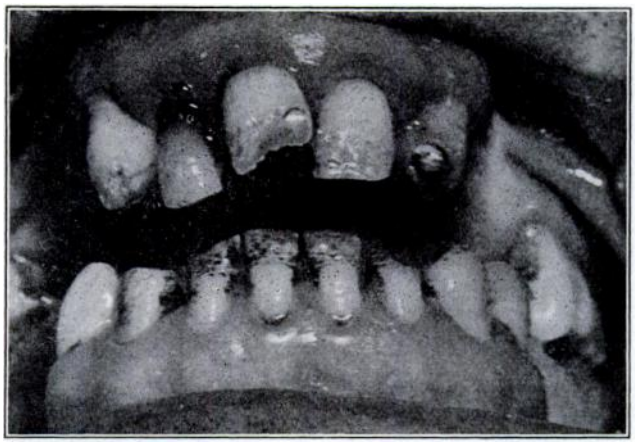

FIG. 3.—H. St—, age 35. Epileptic (imbecile). M.A. 4, 11 years, I.Q. 36. Enamel hypoplasia. Two brothers mentally defective.

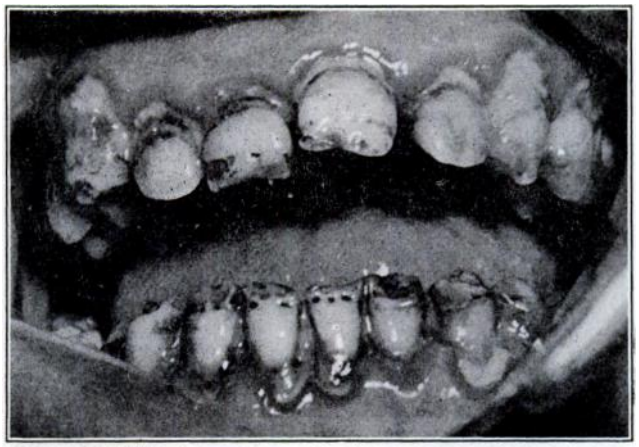

FIG. 4.—E. S—, age 46. Epileptic (imbecile). M. A. 5 years, I.Q. 36. Enamel hypoplasia and dwarfing of the teeth. *Eyes: Positive findings.* Hearing: slightly deaf. Patient is left-handed. One sister mentally defective. Three sisters died in infancy.

tion when they occurred with dental anomalies was striking. In the epileptics coronary opacities were commonest but, in addition, Vogt's axial embryonic cataract (arising at the second month of intra-uterine life) and lamellar cataract were also found and were also associated with dental anomalies.

Among the idiots and imbeciles who were not epileptic, coronary, lamellar and axial cataracts also occurred and were again associated with dental

anomalies. Occasionally, disturbances at two levels in the lens (e.g. Vogt's anterior axial and coronary cataract) were found in the same eye in patients with abnormal teeth. It is, however, obvious from the tables that the associated incidence of lenticular and dental anomalies is highest in the mongols and lowest

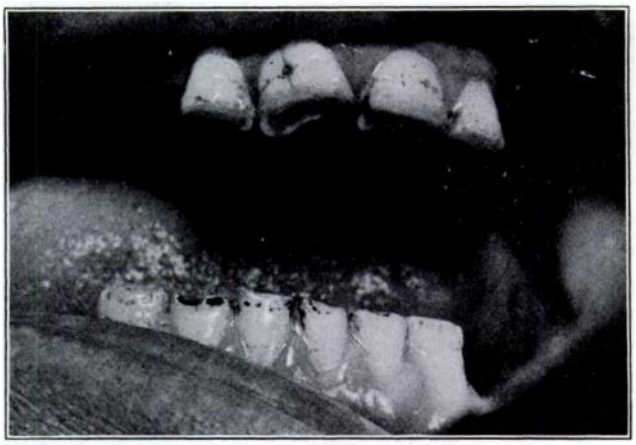

FIG. 5.—C. McC—, age 28. Unclassified mentally deficient patient (imbecile). M.A. 5, 6 years, I. Q. 40. Teeth: Enamel hypoplasia. Eyes: Positive findings. Heart: Mitral stenosis. One sister and six cousins are mentally defective.

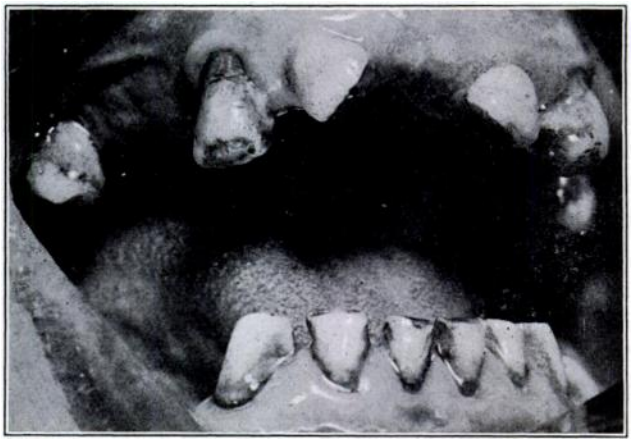

FIG. 6.—J. S—, age 40. Unclassified mental deficient patient (imbecile). M.A. 4, 7 years. I.Q. 33. Teeth: Enamel hypoplasia. Eyes: Positive findings. Two brothers mentally defective.

in the unclassified mental deficient group. This might be expected when one considers the greater structural (skeletal and other mesodermal) anomalies present in mongols than in the other groups.

This association of two ectodermal anomalies (teeth and lens) in individuals with defective cerebral development points to an upset (environmental? genetic?) occurring at an early age. The connection of hypoplastic teeth with

lamellar cataract and the presence of cataract in mongolism has long been known but the present observations extend the principle of lenticulo-dental association in development over a greater range of disorders than was formerly suspected, and afford a possible means of assessing the developmental nature of certain defects, such as epilepsy, which otherwise might have arisen from post-natal environmental causes.

The following tables (I, II, III) show the type of lens opacity and the probable date at which the abnormal lens fibres developed. The fact that in a number of cases with abnormal teeth the opacity did not form until adolescence, does not rule out a genetic cause, since developmental anomalies are not necessarily congenital. In 10 cases evidence of both early ante-natal and adolescent abnormality existed in the same eye. In many cases also, familial defect was apparent; in some the ocular and dental anomaly was accompanied by developmental defect in other organs as well, e.g. deaf-mutism, cardiac lesions, digital anomalies, hemiplegia and cleft palate, all pointing to an early upset of general development. The association of deaf-mutism, cataract and cardiac lesions suggests a disturbance at the second month, but it is interesting that in those cases which showed it the cataract is different in type and not so severe as that found in the rubella syndrome.

Discussion.

The purpose of this survey is to show that various dental and lenticular anomalies are part of a symptom complex, pre-natal in origin.

In addition, an attempt has been made to assess whether these defects are wholly inherited, whether they are acquired foetal lesions, or whether they are due to a summation of both these factors.

The pre-natal origin of the peg-shaped, malformed tooth and of microdontia is undoubted.

Enamel hypoplasia belongs to a different category. It may be due either to pre-natal or post-natal disturbances. Two different types of enamel defects exist, commonly referred to as enamel hypoplasia, although this terminology is not fully satisfactory.

The first is the true hypoplasia of developmental origin, due to an imperfect or aplastic enamel organ resulting from hereditary or early foetal interferences. The other is an enamel lesion caused by disturbance of the calcium metabolism giving rise to hypocalcification effects.

In both, the tooth anomalies are frequently associated with lens defects due to the similarity of origin in both organs (Mann, 1937; Parsons and Stallard, 1942). Such acquired structural enamel defects have been observed in cases of rickets, malnutrition and disturbance of the endocrine function, e.g. parathyroid deficiency (Sarnat and Schour, 1941, 1942; Fish, 1948). However, if post-natal causes are indicted, the diseases have to act over a long period of time and before the termination of the calcification process to produce marked enamel changes. They are usually linked with arrested bone formation.

Recent papers on the aetiology of dental anomalies (Goldman and Bloom, 1949; Boyd, 1943) emphasize that true enamel hypoplasia is primarily linked

TABLE I.—*Leavesden Hospital.—Idiopathic Epileptics*, 100 *Cases*.

Systemic mental disease.	No. of cases.	Sex. Male.	Sex. Female.	Mental age. From	I.Q. From	Tooth changes. Positive.	Tooth changes. Hypoplasia.	Tooth changes. Malformations.	Tooth changes. Microdontia.	Tooth changes. Doubtful.	Tooth changes. Negative.	Other associated congenital lesions. Retarded development.	Other associated congenital lesions. Deaf mutism.	Other associated congenital lesions. Cardiac lesions.	Cases with mental deficiency in family.
Feeble-minded	49	19	30	7	45	13	12	1	1	4	32	14	Deaf, 1	..	Mental deficiency in family, 7. Congenital syphilis, 2. Trauma, 6. Birth premature, 1. Meningitis, 2. Hemiplegia, 3.
Imbeciles	45	20	25	4–6·8	Up to 44	18	17	2	1	2	25	15	Deaf, 3 Deaf & Dumb, 1	..	Mental deficiency in family, 6. Congenital syphilis, 1. Trauma, 3. Birth premature, 2. Meningitis, 2. Hemiplegia, 2.
Idiots	6	2	4	1–3	Up to 19	2	2	..	..	1	3	3	..	..	Trauma, 1.
Total	100	..	..	..	..	33	..	..	..	7	60	..	..	..	

TABLE II.—*Leavesden Hospital.—Unclassified Mentally Deficient Patients.* 105 *Cases.*

Systemic mental disease.	No. of cases.	Sex. Male.	Sex. Female.	Mental age.	I.Q.	Tooth changes. Positive.	Hypoplasia.	Malformations.	Microdontia.	Doubtful.	Negative.	Other associated congenital lesions. Retarded development.	Deaf Mutism.	Cardiac lesions.	Cases with mental deficiency in family.
Feeble-minded	38	15	23	From 7	From 45	3	3	1	..	..	35	1	..	1	Mental deficiency in family, 5. Meningitis, 3. Trauma, 2.
Imbeciles	57	32	25	4–6·8	Up to 44	9	9	2	..	3	45	13	Deaf, 1	1	Mental deficiency in family, 5. Birth premature, 3. Hemiplegia, 1. Trauma, 1.
Idiots	8	5	3	1–3	Up to 19	2	2	..	..	1	5	2	..	..	..
Cretin	2	1	1	..	..	..	..	..	..	..	2	..	..	..	..
Total	105	..	..	..	..	14	..	..	..	4	87	..	..	..	

TABLE III.—*Leavesden Hospital.—Mongols, 25 Cases.*

Systemic mental disease.	No. of cases.	Sex. Male.	Sex. Female.	Mental age.	I.Q.	Tooth changes. Positive.	Hypoplasia.	Malformations.	Microdontia.	Doubtful.	Negative.	Other associated lesions. Retarded development.	Deaf Mutism.	Cardiac lesions.	Cases with mental deficiency in family.
Feeble-minded	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Imbeciles	18	4	14	4–6·8	Up to 44	13	3	8	5	1	4	16	..	1	Mental deficiency in family, 4.
Idiots.	7	4	3	1–3	Up to 19	4	..	3	1	1	2	5	..	1	..
Total	25	8	17	..	..	17	..	..	..	2	6	..	..	..	..

TABLE IV.—*Fountain Hospital.—Idiopathic Epileptics, 15 Cases.*

Systemic mental disease	No. of cases.	Sex. Male.	Sex. Female.	Mental age.	I.Q.	Tooth changes. Positive.	Hypoplasia.	Malformations.	Microdontia.	Doubtful.	Negative.	Other associated congenital lesions. Retarded development.	Deaf Mutism.	Cardiac lesions.	Notes on patients' history and cases with mental deficiency in family.
A. Feeble minded	1	..	1	From 7	From 45	..	..	..	..	..	1	..	..	..	Mental deficiency in family, 1.
B. Imbeciles	12	..	12	4–6·8	Up to 45	3	3	1	..	1	8	2	..	..	Birth premature, 3. Mental deficiency in family, 5.
C. Idiots.	2	..	2	1–3	Up to 19	..	..	..	..	..	2	..	..	..	Cannot speak, 1.
Total	15	..	15	..	..	3	..	..	..	1	11	..	..	..	..

TABLE V.—*Fountain Hospital.—Unclassified Mentally Deficient Patients.* 61 *Cases.*

Systemic mental disease	No. of cases.	Sex. Male.	Sex. Female.	Mental age.	I.Q.	Tooth changes. Positive.	Hypoplasia.	Malformations.	Microdontia.	Doubtful.	Negative.	Other associated congenital lesions. Retarded development.	Deaf Mutism.	Cardiac lesions.	Notes on patients' history and cases with mental deficiency in family.
A. Feeble-minded	14	..	14	From 7	From 45	2	2	..	..	..	12	2	..	..	Mental deficiency in family, 6.
B. Imbeciles	42	1	41	4–6·8	Up to 45	5	5	1	..	1	36	Microcephalic, 1 1	..	..	Mental deficiency in family, 7. Congenital syphilis, 1.
C. Idiots	5	..	5	1–3	Up to 19	..	..	..	..	..	5	..	..	..	Congenital syphilis, 1.
Total	61	1	60	..	..	7	..	..	..	1	53	..	..	..	..

TABLE VI.—*Fountain Hospital.—Mongols,* 13 *Cases.*

Sytemic mental disease.	No. of cases.	Sex. Male.	Sex. Female.	Mental age.	I.Q.	Tooth changes. Positive.	Hypoplasia.	Malformations.	Microdontia.	Doubtful.	Negative.	Other associated congenital lesions. Retarded development.	Deaf Mutism.	Cardiac lesions.	Notes on patients' history and cases with mental deficiency in family.
A. Feeble-minded	..	..	..	From 7	From 45	..	..	..	..	..	..	..	..	..	..
B. Imbeciles.	12	1	11	4–6·8	Up to 45	9	..	8	3	..	3	4	..	..	..
C. Idiots	1	..	1	1–3	Up to 19	1	..	1	..	..	..	1	..	..	..
Total	13	1	12	..	..	10	..	..	..	..	3	..	..	..	..

with either a faulty development in the enamel matrix or imperfect tooth primordia. In addition, Cockayne (1933) and Fish (1948) mention instances of enamel hypoplasia due to a primary ectodermal lesion. The reasons for assuming that the structural enamel lesions reported in this paper are due to pre-natal interference are:

1. The incidence is decidedly greater in the mental defectives than in the normal population.

2. There is a definite correlation in the incidence between these enamel defects and other proved congenital anomalies.

3. The most important associated clinical defects are in the lens. These, which are accepted as developmental, suggest that the same aetiological factors are acting to produce the dental lesions.

4. The lesions in the enamel were more marked than would be likely to occur with post-natal causes.

5. No other stigmata or clinical symptoms were found which could have been ascribed to post-natal influences.

6. The presence of dental lesions with developmental anomalies and mental deficiency is consistent with the pattern of other well-defined congenital diseases. Some of the questions involved will be better understood if the function of the embryonic layer in the formation of the tooth primordia is first briefly considered. The enamel of the tooth is a product of the ectoderm which starts to differentiate into the dental lamina in the 6 to 7 weeks' embryo. This is the time at which the ectodermal lining of the mouth cavity grows down into the mesenchyme and forms the enamel organ. From this enamel organ the decidous teeth are first produced, but the same organ also produces the permanent teeth, and these are thus part of the primary dental lamina. This primordium already bears the inherent stigmata of hereditary or early environmental influences and determines, to a large degree, shape and structure of both dentitions. It must be remembered in this connection that not only the enamel but also nervous tissue and lens are of the same ectodermal derivation.

In cases where there is an interference with the development three different types of dental disorders may appear:

1. Anodontia.

2. Peg-shaped tooth and microdontia.

3. Enamel hypoplasia. This is a condition where defective enamel prisms will be added to the unimpaired tissue. This defect persists and forms the hypoplastic lesion (Mellanby, M., 1934; and Mellanby, H., 1941). Any such lesion of the enamel cannot be repaired at later stages of embryonic or post-natal development. Similarly, lenticular embryonic defects are irreversible (Mann, 1937; Bellows, 1944). It has been a subject of controversy whether the clinically apparent dental defects are due to genetic causes or are due to interference by environmental influences acting before birth or shortly after.

Any one of these causes may affect the tooth in its developmental state. A lesion of the ectoderm or deficient ameloblasts can constitute a cause for the dental anomalies. Similar changes will appear if the cells are subjected to inflammation during the period of maximum cellular activity. Since the

primordiae of the teeth are laid down at different periods of foetal life, the congenital manifestations will vary in degree and location according to cause and time, but such changes will be bilateral and identical, while other groups of teeth may completely be unaffected.

In spite of the importance of both genetic and acquired factors in the aetiology of mental disease, the dental manifestations have been little studied. The dental lesions in the cases reported are clinically allied with mental diseases which are regarded as pre-natal. In a significant number of cases other defects of undoubted congenital origin besides the lens changes are present. The associated complications are cleft palate, hare-lip, webbing of fingers and toes, brachycephaly, epicanthus and deafness. All these cannot be regarded as incidental complications.

The high incidence of associated dental and lenticular anomalies are very suggestive of a common aetiological factor or factors.

The clinical examination is, with all its limitations, one way to approach the problem of whether the tooth defects are due to post-natal environmental influences or whether they are congenital. The findings in this paper provide important evidence of the latter. Among 51 idiopathic epileptics in Leavesden Hospital (Tables VIIA, B, C, and Table X) there were 20 with both dental and lenticular lesions. Among these 51 patients, 9 showed other congenital malformations or lesions. Although the family history was available only in a few cases, 7 patients in this group had records of mental deficiency in the family. Of the 19 mongols (Tables VIIIA, B, and Table XI), 14 cases had both dental and lenticular lesions. Four patients in this category had records of mental deficiency in the family. Seven of these 14 cases exhibited other congenital malformations such as webbed fingers or toes (one case), congenital heart defects (2 cases), and epicanthus and brachycephaly (one case), and 3 others showed minor congenital changes such as high palate or a fissured tongue. Of 5 unclassified mentally deficient patients (Tables IXA, B, and Table XII) with positive findings in teeth and eyes, 2 gave histories of mental deficiency in the family. One (J. S—) had an epileptic brother in the same hospital, another (Th. B—) had an epileptic father and a feeble-minded sister. A third case (Ch. McC—), with dental lesions and a slight anomaly of the retinal disc (inferior crescent), had one mentally defective sister and 6 mentally defective cousins. There were no data on the family history of the other 3 cases. One of these 6 patients (E. M—), with no family history or records on the sibship, was slightly deaf and had a high palate.

Of the 4 cases with negative findings in teeth and eyes, 2 patients had a history of trauma and one had had encephalitic lethargica which was thought to be the possible cause of the mental condition. An analysis of both these positive and negative findings shows that dental disorders are much more prevalent among these mental patients than among the general population. Bruckner (1943), cited by Stones (1948), gives the incidence of hypoplasia in the permanent teeth among the general population at 0·7 per cent. A higher percentage (5 per cent.) has been found in Chicago schoolchildren by Greenwald (quoted by Sarnat and Schour, 1942). The absence of teeth (anodontia) and certain types of tooth anomalies, such as the malformation of shape (haplodont

TABLE VII.—*Leavesden Hospital.—Epileptics (Feeble-minded).*

Name.	Age.	Sex.	Ment. age.	I.Q.	Congenital defects in patients.												Notes on patients' history and mental def. in family.
					Tooth changes.						Eye changes.			Other congenital conditions.			
					Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Positive.	Negative.	Doubtful.	Deafmutism.	Cardiac lesions.	Other cong. malformations.	
1. F. D—	30	F.	9	..	+	+	−	−	−	−	+	−	−	..	..	..	Mother and cousin on maternal side ment. def.
2. P. P—	26	F.	7·7	55	+	−	+	−	−	−	−	+	−	..	..	Cleft palate. Very thin and long fingers and toes.	Birth premature.
3. M. D—	37	F.	10·8	..	−	−	−	−	−	+	+	−	−	..	..	..	
4. A. Y—	19	F.	..	..	−	−	−	−	−	+	−	+	−	..	..	..	"Stroke," meningitis when 9 months old.
5. E. A. P—	28	M.	7·3	52	−	−	−	−	−	+	−	+	−	..	..	..	Def. observed at 16 years of age. Pat. knocked down by car when 12 years old.
6. J. R. W—	29	M.	..	..	−	−	−	−	−	+	−	+	−	..	..	..	Def. observed at 7 years of age. Brother died at mental hosp.
7. A. E. H—	37	M.	7·5	..	−	−	−	−	−	+	−	+	−	..	..	..	Trauma. Fall when 3 years old. Def. observed when 5 years old.
8. F. M—	41	M.	8·6	61	−	−	−	−	−	+	−	+	−	..	..	..	Def. observed at 4 years of age.
9. T. W. B—	27	M.	10·3	73	−	−	−	−	−	+	+	−	−	..	..	..	Trauma. Knocked down by car when 5 years old. Unconscious.
10. J. H—	23	F.	..	..	−	−	−	−	−	+	−	+	−	..	..	Right hemiparesis	Noted at school age.
11. W. M—	31	F.	7·3	52	−	−	−	−	−	+	−	+	−	..	+	Slightly deaf	..
12. D. A. G—	33	F.	9·6	66	−	−	−	−	−	+	−	+	−	..	..	Slightly deaf	Def. observed when 10 years old. Sister ment. def.

TABLE VIIA.—*Leavesden Hospital.—Epileptics (Feeble-minded)—contd.*

Name.	Age.	Sex.	Ment. age.	I.Q.	Congenital defects in patients. Tooth changes. Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Eye changes. Positive.	Negative.	Doubtful.	Other congenital conditions. Deafmutism.	Cardiac lesions.	Other cong. malformations.	Notes on patients' history and mental def. in family.
13. M. D. B—	26	F.	7	50	+	+	—	—	—	—	+	—	—	..	..	..	..
14. J. G. G—	28	F.	9·6	69	—	—	—	—	+	—	—	+	—	..	..	..	Meningitis when 13 months old.
15. H. A. F—	42	M.	..	..	+	+	—	—	—	—	+	—	—	..	..	Palate high. Internal strabismus	..
16. F. A—	47	F.	7·8	..	+	—	—	—	—	—	—	+	—	..	..	Hutchinson teeth in maxilla	W.R. +. Parallel Tests ++++.
17. N. L—	34	F.	10·8	77	—	—	—	—	—	+	—	+	—	..	..	..	..
18. W. G. R—	34	M.	8	57	+	+	+	—	—	—	—	+	—	..	..	Fifth finger on left hand shorter than right	..
19. C. S—	47	F.	8·5	61	+	+	—	—	—	—	+	—	—	..	..	..	..
20. E. B—	45	F.	6·5	46	+	+	—	—	—	—	—	+	—	..	..	..	..
21. G. E. W—	44	F.	6·8	49	—	—	—	—	—	+	—	+	—	..	..	Left facial palsy with wasting	Paralysis at the age of 2.
22. H. E. H—	32	F.	10·2	73	—	—	—	—	—	+	—	+	—	..	..		Spastic paraplegia (brain injury). Def. noted at age of 13.
23. G. W—	25	F.	7·2	51	—	—	—	—	—	+	—	+	—	..	..	..	..
24. E. E. W—	27	F.	8·9	62	—	—	—	—	—	+	—	+	—	..	..	..	Trauma: knocked down by car when 5.
25. A. D—	25	M.	8	57	—	—	—	—	—	+	—	+	—	..	..	..	..
26. J. R. H—	41	M.	7·1	56	+	+	—	+	—	—	+	—	—	..	..	..	..
27. G. M—	45	M.	8	57	+	+	—	—	—	—	+	—	—	..	..	..	Father epileptic.
28. J. A. H—	28	F.	..	..	+	+	—	—	-	—	+	—	—	..	..	..	Ment. def. on the paternal side. W.R., positive. Parallel negative.

Total: 28 cases.
Males: 10.
Females: 18

Tooth changes:
Positive: 11 cases.
Negative: 16 cases.
Doubtful: 1 case.

TABLE VIIB.—*Leavesden Hospital.—Epileptics (Imbeciles).*

Name.	Age.	Sex.	Ment. age.	I.Q.	Congenital defects in patients. Tooth changes. Malformation. Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Eye changes. Positive.	Negative.	Doubtful.	Other congenital conditions. Deaf mutism.	Cardiac lesions.	Other cong. malformations.	Notes on patients' history and mental def. in family.
1. C. D—	25	M.	Tests inapplicable.		+	+	−	−	−	−	+	−	−	+	..	High palate	Mother, 2 brothers, one sister ment. def. One brother and sister deaf and dumb.
2. A. M—	64	M.	4·7	32	+	+	−	−	−	−	+	−	−	..	..	Defective speech	..
3. R. W—	45	F.	5·7	40	+	+	−	−	−	−	+	−	−	..	..	..	Father deaf. Paternal Grandfather ment. def.
4. E. S—	47	F.	5	36	+	+	+	−	−	−	+	−	−	..	..	Slight deafness Right hemiplegia	Sister ment. def.
5. F. S—	37	F.	Tests inapplicable		+	+	−	+	−	−	+	Nil. Developmental syphilitic		..	..	..	..
6. E. A. P—	43	F.	5·6	40	+	+	−	−	−	−	+	−	−	..	Mitral stenosis	Left hemiplegia	..
7. J. G. B—	37	M.	..	..	−	−	−	−	+	−	+	−	−	..	..	..	History of insanity on the maternal side.
8. E. J. H—	45	F.	4·3	32	+	+	−	−	−	−	+	−	−	..	..	Deaf	..
9. B. S—	44	F.	5	37	+	+	−	−	−	−	+	−	−	..	..	Palate high. Little fingers abducted from midline	..
10. H. R. H—	28	M.	5·9	41	−	−	−	−	−	+	−	+	−	..	..	Palate high	..
11. T. W. C—	40	M.	..	..	+	+	−	−	−	−	−	+	−	..	..	Left hemiplegia	Trauma (accident when one year old).
12. H. S—	35	M.	4·11.	36	+	+	−	−	−	−	−	+	−	..	..	Palate high Adherent lobes	One brother ment. def.
13. N. S—	43	F	3·5	25	+	+	−	−	−	−	−	+	−	..	..	..	Meanacke +++. Trauma and meningitis when 5 months old.
14. P. M. W—	28	F.	Tests inapplicable		−	−	−	−	−	+	−	+	−	..	..	..	Mother suffered motor accident in third month of pregnancy.
15. W. S—	43	F.	5·1	37	−	−	−	−	+	−	−	+	−	..	..	Left hemiplegia	Birth premature (6 months).
16. R. B—	26	F.	3·8	27	+	+	−	−	−	−	+	−	−	..	..	Left hemiplegia	Grandfather ment. def.
17. R. M. D—	25	F.	5	36	−	−	−	−	+	−	+	−	−	..	..	Right hemiplegia	..
18. J. B—	47	M.	4·4	31	+	+	−	−	−	−	+	−	−	..	..	..	..
19. C. W. L—	35	M.	5·6	39	+	+	−	−	−	−	+	−	−	..	..	..	Mother had a fall during pregnancy. Birth difficult and prolonged.
20. A. M. F—	46	F.	6	..	−	−	−	−	−	+	−	+	−	..	..	Slight epicanthus	Def. noted at school age. Mother ment. def.

TABLE VIIc.—*Leavesden Hospital.—Epileptics (Idiots).*

Name.	Age.	Sex.	I.Q.	Ment. age.	Congenital defects in patients.											Notes on patients' history and mental def. in family.
					Tooth changes.						Eye changes.			Other congenital conditions.		
					Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Positive.	Negative.	Doubtful.	Deaf mutism.	Cardiac lesions.	Other cong. malformations.	
1. J. R—	41	M.	18	2·5	−	−	−	−	+	−	−	+	−	..	..	..	..
2. M. K—	46	M.	20	2·9	+	+	−	−	−	−	+	−	−	..	..	Mitral stenosis	..
3. I. G—	31	F.	19	2·7	+	+	−	−	−	−	+	−	−	..	..	Palate high	Deficiency was observed from infancy.

TABLE VIIIa.—*Leavesden Hospital.—Mongols (Imbeciles).*

Name.	Age.	Sex.	Ment. age.	I.Q.	Congenital defects in patients.											Notes on patients' history and mental def. in family.
					Tooth changes.						Eye changes.			Other congenital conditions.		
					Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Positive.	Negative.	Doubtful.	Deaf mutism.	Cardiac lesions.	Other cong. malformations.	
1. H. F—	27	M.	32	4·6	+	−	+	+	−	−	+	−	−	..	..	Palate high. Speech mongolian	One miscarriage, other sibs normal.
2. G. L. H—	33	F.	37	5·2	+	+	+	−	−	−	+	−	−	..	..	Tongue mongolian	Pat. is the fourth child Three others normal.
3. C. C—	24	F.	27	3·8	+	−	−	+	−	−	+	−	−	..	..	..	Cousin, imbecile. Mother, defective speech.
4. F. H—	41	F.	33	4·7	+	−	−	+	−	−	+	−	−	..	..	Brachycephaly epicanthus	..
5. M. L. P—	27	F.	38	5·3	+	−	−	+	−	−	+	−	−	..	..	..	Premature birth.
6. F. R—	31	F.	27	3·8	+	+	+	−	−	−	+	−	−	..	..	Defective speech Palate high. Tongue mongolian	Sister imbecile (dead): Aunt had epilepsy. Pat. last of 9 pregnancies.
7. P. N. S—	21	F.	39	5·5	+	−	+	+	−	−	−	−	+	..	..	..	Defective is the second child of a family of 3; the other 2 are normal.
8. J. H—	27	F.	..	..	+	−	+	+	−	−	+	−	−	..	..	Tongue fissured	..
9. H. R—	27	F.	32	4·5	+	−	−	+	−	−	+	−	−	..	..	..	Maternal grandfather died in mental hospital.
10. A. E. T—	37	M.	..	..	+	+	+	−	−	−	+	−	−	..	..	..	Pat. is the first in a family of 2. Father's brother was a mental defective. Father died in diabetic coma. Pat. suffered from *petit mal* in childhood.
11. J. H. D—	32	M.	3·8	26	+	−	+	+	−	−	−	+	−	..	..	Palate high	..
12. D. K—	40	F.	..	..	+	−	−	+	−	−	+	−	−	..	..	Epicanthus right and left. Speech defective	..
13. L. E. K—	25	M.	3·6	25	−	−	−	−	−	+	−	+	−	..	..	Palate high. Tongue mongolian	..
14. F. M. S—	43	F.	2·8	20	−	−	−	−	−	+	−	+	−	..	..	..	..
15. E. M. C—	28	F.	3·8	26	+	+	+	−	−	−	+	−	−	..	+	Palate high. Tongue mongolian	..

TABLE VIIIB.—*Leavesden Hospital.—Mongols (Idiots).*

Name.	Age.	Sex.	Ment. age.	I.Q.	Congenital defects in patients. Tooth changes. Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Eye changes. Positive.	Negative.	Doubtful.	Other congenital conditions. Deaf mutism.	Cardiac lesions.	Other cong. malformations.	Notes on patients' history and mental def. in family.
1. G. G. S—	35	M.	17	2·2	+	−	+	−	−	−	+	−	−	..	..	Palate high. Tongue mongolian	..
2. A. G—	23	F.	..	..	+	−	+	−	−	−	+	−	−	..	+	..	..
3. F. M—	43	M.	18	2·6	+	+	−	−	−	−	+	−	−	..	..	Webbed fingers on left hand, webbed toes on both feet. Skin not very much pigmented	..
4. B. L. G—	26	F.	..	..	−	−	−	−	+	−	−	+	−	..	..	..	Birth premature.

TABLE IXA.—*Leavesden Hospital.—Unclassified Mental Deficient Patients (Feeble-minded).*

Name.	Age.	Sex.	Ment. age.	I.Q.	Congenital defects in patients. Tooth changes. Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Eye changes. Positive.	Negative.	Doubtful.	Other congenital conditions. Deaf mutism.	Cardiac lesions.	Other cong. malformations.	Notes on patients' history and mental def. in family.
1. F. O—	41	M.	11·8	84	−	−	−	−	−	+	−	+	−	..	..	..	Encephalitis lethargica when 12 years old; up to that date bright and normal.
2. E. H—	40	M.	10·2	72	−	−	−	−	−	+	−	+	−	..	..	..	Trauma in history.
3. D. R. J—	24	F.	6·5	46	+	−	+	−	−	−	−	+	−	..	..	..	Mother had fall during pregnancy. One brother feeble-minded. Age of talking: 4 years.
4. T. B—	28	M.	7·1	..	+	+	−	−	−	−	+	−	−	..	...	..	Father epileptic. Sister feeble-minded.

TABLE IXB.—*Leavesden Hospital.—Unclassified Mental Deficient Patients (Imbeciles).*

Name.	Age.	Sex.	Ment. age.	I.Q.	Congenital defects in patients.												Notes on patients' history and mental def. in family.
					Tooth changes.						Eye changes.			Other congenital conditions.			
					Positive.	Hypoplasia.	Malformation.	Microdontia.	Doubtful.	Negative.	Positive.	Negative.	Doubtful.	Deaf mutism.	Cardiac lesions.	Other cong. malformations.	
1. J. S—	41	M.	4·7	33	+	+	−	+ (slight)	−	−	+	−	−	−	−	..	One brother feeble-minded and epileptic, another brother ment. def.
2. E. M—	41	M.	5·2	37	+	+	+	−	−	−	+	−	−	..	..	Hearing impaired. Palate high	..
3. F. B—	47	M.	4·3	31	+	+	−	−	−	−	−	+	−	..	..	..	Mother feeble-minded, father feeble-minded(?) Twin, other child died at 6 months.
4. B. M. D—	26	F.	5·6	40	−	−	−	−	−	+	−	+	−	..	..	Palate high	Parturition: instruments. Mother had a fall at third month of pregnancy. Maternal cousin: imbecile.
5. G. M. M—	40	F.	5·7	41	−	−	−	−	−	+	−	+	−	..	..	Cretin	..
6. C. S. Mc—	28	M.	5·6	40	+	+	−	−	−	−	−	+	−	..	..	Inf. crescent, left disc (congenital). Enlargment of sternal end of right clavicle	Sister and 6 cousins.
7. H. B—	45	M.	5·1	37	+	+	−	−	−	−	+	−	−	..	..	Speech defective	..
8. W. A—	32	M.	very low. Idiot		+	+	−	−	−	−	+	−	−	..	..	Physique small. Idiot	Endocrine: sexually immature. No pubic or axill. hair.
9. T. S—	35	M.	Under one year. Idiot		+	+ (slight)	−	−	−	−	−	−	+	..	..	..	Birth premature.

TABLE X.—*Leavesden Hospital.—Table of Associated Incidence of Dental and Lenticular Malformations.*
Idiopathic Epileptics: 51 Cases.

		Eyes.			
		Positive.	Doubtful.	Negative.	Total.
Teeth	Positive	20	..	7	27
	Doubtful	2	..	3	5
	Negative	2	..	17	19
	Total	24	..	27	51

TABLE XI.—*Leavesden Hospital.—Table of Associated Incidence of Dental and Lenticular Malformations.*

Mongols: 19 Cases.

		Eyes.			
		Positive.	Doubtful.	Negative.	Total.
Teeth	Positive	14	1	1	16
	Doubtful.	..	..	1	1
	Negative	..	..	2	2
	Total	14	1	4	19

TABLE XII.—*Leavesden Hospital.—Table of Associated Incidence of Dental and Lenticular Malformations.*

Unclassified Mental Deficient Patients: 13 Cases.

		Eyes.			
		Positive.	Doubtful.	Negative.	Total.
Teeth	Positive	5	1	3	9
	Doubtful	..	..	..	..
	Negative	..	..	4	4
	Total	5	1	7	13

tooth), found in mongols, are most unusual and rarely encountered in normal people. These lesions are essentially the result of early dental maldevelopment, which agrees with Penrose's view (1934), that mongolism is a condition of abnormal or arrested development.

Peg-shaped or haplodont teeth are examples of primitive teeth in non-human dentitions from which the crowns of the normal teeth in men have gradually been formed (Keith, 1948). They are an indication of an arrest of growth during the transitional stage of development. Anodontia is a manifestation of an even more severe developmental interference with the ectodermal layer (Thomas, 1939).

Consideration of these cases suggests an aetiological relationship for the dental and other congenital disorders and the mental disease.

When one considers congenital syphilis and maternal rubella, and their effects on the tooth structures, it is clear that congenital lesions need not necessarily be genetically determined. In these conditions the cause of the abnormalities is clearly due to pre-natal environmental factors. On the other hand, in hereditary congenital lesions the genetical causes are the only deter-

TABLE XIIIA.—*Epileptics. 51 Examined. 23 with Eye Changes.*

Name.	Nature of lenticular defect.	Probable time.	Remarks.	Nature of dental lesion.
C. D.—	Diffuse opacity in posterior cortex and sutures of adult nucleus	From birth onwards	Small, persistent hyaloid artery, also patient is deaf mute with "Gothic" palate. Teeth abnormal	Hypoplastic lesions in grooves.
A. M—	R.: opaque dislocated lens. L.: Vogt's ant. axial cataract.	Post-natal trauma. Second month ante-natal.	Teeth abnormal	Hypoplastic lesions in shallow pits and grooves.
R. W—	Dot-like opacities in posterior cortex	Early post-natal	Anomaly very slight. Teeth abnormal	Hypoplastic lesions.
F. D—	Arcuate opacities and Vogt's axial cataract	Second month ante-natal and at birth	Teeth abnormal	Hypoplastic lesions in pitted grooves.
E. S—	Lamellar cataract	Third month ante-natal to third month post-natal	ditto	Hypoplastic lesions at the incisal parts, some teeth dwarfed.
F. S—	Secondary cataract from syphilitic iritis	..	,,	Hypoplastic lesions in pits, malformed teeth, microdontia.
E. P—	Dot-like opacities in cortex	Post-natal, early	,,	Hypoplastic lesions.
J. B—	Single layer of dotted cataract	At birth	Teeth normal	..
T. W. B—	Coronary cataract	Adolescence	ditto	..
M. D—	Coronary and blue dot cataract	,,	,,	..
J. H—	Punctate opacities in posterior cortex	Birth	Teeth abnormal	Hypoplastic lesions near the cervical parts.

TABLE XIIIB.—*Epileptics.* 51 *Examined.* 23 *with Eye Changes.*

Name.	Nature of lenticular defect	Probable time.	Remarks.	Nature of dental lesion.
M. B—	Very few posterior punctate opacities	Birth	Teeth abnormal	Hypoplastic lesions, very thin layer of enamel at other parts.
H. A. F—	Punctate cortical opacities	Just post-natal	ditto	Hypoplastic lesions in horizontal grooves.
M. K—	Opacities on surface of adult nucleus	Late adolescence	,,	Dense hypoplastic lesions of the enamel.
I. G—	Vogt's axial cataract	Second month ante-natal	,,	Hypoplastic lesions in pits and grooves.
E. J. H—	Coronary cataract	Adolescence	,,	Hypoplastic lesions of the enamel, not very marked.
B. S—	Lamellar cataract in two separate zones	Sixth month ante-natal and adolescence	,,	Enamel hypoplasia, not very marked. High palate.
R. B—	Arcuate type of cataract in a single layer	Eighth month ante-natal	Teeth hypoplastic	Enamel hypoplasia in grooves near the incisal edges.
R. D—	Secondary cataract	Post-natal	Teeth inconclusive. Eye condition is secondary to iritis	..
J. B—	Vogt's axial cataract	Second month ante-natal	Also shows congenital anomaly of optic disc. Teeth inconclusive	Thin hypoplastic lesions of the enamel.
C. L—	Dot-like opacities in foetal nucleus	Third to eighth month ante-natal	Teeth slightly abnormal	Enamel hypoplasia in grooves near the incisal edges.
J. R. H—	Punctate opacity in posterior foetal nucleus	End of foetal life	Teeth abnormal	Small hypoplastic grooves and microdontia.
G. M—	Small lamellar cataract	Second to sixth month ante-natal	ditto	Enamel hypoplasia in horizontal grooves.

TABLE XIV.—*Feeble-minded and Imbeciles.* 13 *Examined.* 5 *with Eye Changes.*

Name.	Nature of lenticular defect.	Probable time.	Remarks.	Nature of dental lesion.
J. S—	Flake opacities	Between 8 months ante-natal and 6 months post-natal	Teeth abnormal	Densely pitted hypoplastic grooves. Teeth slightly dwarfed.
T. B—	Dust-like opacities in foetal nucleus and blue dot and coronary	Before fifth month and during adolescence	ditto	Marked hypoplastic horizontal grooves.
E. M—	Coronary cataract and Vogt's axial cataract	Second month ante-natal and during adolescence	,,	Marked hypoplastic defects.
H. B—	Lamellar cataract	Three months ante-natal	,,	Hypoplasia in grooves and pits.
W. A—	Coronary type. Very slight	Adolescence	,,	Hypoplasia.

TABLE XV.—*Mongols.* 19 *Examined.* 14 *with Eye Changes.*

Name.	Nature of lenticular defect.	Probable time.	Remarks.	Nature of dental lesion.
G. S—	Subcapsular	Late adolescence	Teeth abnormal	Malformed peg-shaped teeth, with dented incisal edges.
H. F—	Coronary cataract	Adolescence	ditto	Malformed peg-shaped teeth, with serrated incisal edges.
G. L. H—	ditto	,,	,,	Hypoplastic lesions and partial microdontia.
C. C—	Cortical opacities slight	Late adolescence	,,	Severe microdontia.
F. H—	Coronary and arcuate opacity, severe	Last months of foetal life and during adolescence	,,	Microdontia.
M. L. P—	Suture and dot cataract	Fifth month ante-natal and during adolescence	,, (microdontia)	Severe microdontia.
D. K—	Coronary cataract, severe	Adolescence	Teeth abnormal (microdontia)	ditto
A. G—	Coronary and suture cataract	Fifth month ante-natal and adolescence	Teeth grossly abnormal	Grossly malformed peg-shaped teeth.
J. H—	Severe coronary cataract	Adolescence	Teeth abnormal	Microdontia, defective enamel.
H. R—	Subcapsular opacities	Late adolescence	ditto	Severe microdontia.
A. E. T—	Coronary opacities	Adolescence	,,	Hypoplastic defects, peg-shaped canines.
F. R—	Severe coronary cataract	,,	,,	Malformed peg-shaped teeth, with defective enamel.
F. McC—	Slight coronary cataract	Late adolescence	,,	Hypoplastic lesions in pits.
E. M. C—	Suture and dot cataract	Fifth month ante-natal and during adolescence	,,	Peg-shaped malformed teeth, with jagged incisal edges.

minant of the disease. A good example is the hereditary ectodermal dysplasia of the anhidrotic type. The condition is generally regarded as due to a recessively inherited gene, and involvement of the teeth is an example of a dental anomaly of such origin. The argument that teeth are under genetic control is supported by Burks (1938), Snyder (1941), and Gates (1946), who discuss various dental disorders of genetic aetiology.

Benda emphasizes that malformations of the central nervous system are usually associated with other stigmata of arrested development. The evidence from the data presented establishes a linkage between the causes of the endogenous mental deficiency and associated developmental lesions. There is unanimity of opinion as to the hereditary character of feeble-mindedness. Regarding idiopathic epilepsy, the constitutional factors in the causation of this disorder are generally considered to be of great importance. Lennox (1947) concludes that predisposition to epilepsy, rather than epilepsy itself, is inherited, and that probably in most patients genetic and acquired factors are instrumental in producing the epileptic condition.

Mongolism comes into a third category, in which the disease is only partly determined by heredity (Penrose, 1934; Halperin, 1946). Other causes, such as the age of the mother and congenital endocrine dysfunction, are mentioned (Benda, 1947). Recently, Ingalls and Gordon (1947) have suggested that uterine bleeding in the early months of gestation may constitute an environmental condition leading to mongolism.

It is well known that mongols exhibit the largest variety of malformations among the mental defectives. Organs of all three derivatives are affected.

As regards the dentition, the skeletal deformities such as the high, arched palate, the cleft palate and the small jaws with the resulting malocclusion can be ascribed to disturbances in the mesodermal layer.

The other, more severe structural disorders of the dental tissue itself, the peg-shaped form, the microdontia and the anodontia are likely to be predominantly conditioned by the same causes which produce the physical characteristics of the skin, lens and hair. They are all indicative of a defective ectodermal layer, or of an impairment to its normal growth.

It is of interest to note that the front, or oral, part of the tongue is the seat of the typical changes in mongols. This part is covered with a mucosa of ectodermal origin. The inference from these observations is that the dental lesions are aetiologically allied with the other anomalies.

There are transitional forms of dental malformations to be found in these cases, from the near-normal to the grossly abnormal. The teeth in the mongols present the most pronounced dental defects in all three groups.

In mongols the nature and pathology of the systemic condition account for a dental manifestation (microdontia, peg-shaped teeth, anodontia and hypoplasia), which seems to be induced by developmental interference. The idiopathic epileptics and imbeciles show milder types of anomalies. The dental lesions seen in idiopathic epileptics and imbeciles are mostly hypoplasia of the enamel, which, however, in itself, is no less a significant sign of defective development.

The stage of development of the afflicted organ makes it more vulnerable

at one period of gestation than at the other. This fact provides a possible explanation for the presence or absence of anomalies, whose production may be attained or supressed in one stage or the other. Experimental investigations have shown (von Bahr, 1936; Mellanby, 1941) that the onset of environmental causes can only take effect before the differentiation and calcification of the tooth organ is terminated. The combination and interrelation of these factors explain the occurrence, types and degrees of the dental lesions.

A conclusive answer to the various questions involved will necessitate a study of the histological and radiological appearance of teeth and jaws, as well as the clinical examination, since the microscopic examination may disclose even those congenital anomalies which are subclinical but, nevertheless, significant for the pathology. In addition, the other ectodermal derivatives have to be included in such an investigation, since variable conditions occur in skin, hair and nails. They may also reveal manifestations of an inhibited or arrested development (Cockayne, 1933).

To test the theory of a genetic tendency, it may be of importance to study the electroencephalogram in cases with both dental and lenticular lesions. It is conceivable that epileptics and mentally deficient patients with such lesions may have electroencephalographic patterns differing in some way from the tracings obtained in cases without such associated disorders (Lennox and Gibbs, 1944; Lennox, 1947). It is hoped to submit a series of patients for such an investigation later on.

Furthermore, the examination of the near relatives is essential, in order to see whether an hereditary trait can be found in this pathology (Penrose, 1934; Benda, 1946; Halperin, 1946). The extension of the present investigation in these two directions would be an aid in the evaluation and interpretation of the underlying causes of the dental anomalies we have been considering.

Summary.

An investigation has been made on the occurrence of various disorders of the teeth and the lens in mental defectives. In order to ascertain the incidence of developmental malformations, 319 mental cases were examined clinically. Of these, 83 underwent slit-lamp examination of the lenses.

The structural pattern of the teeth was affected in 84 cases. The incidence of dental disorders in these patients is thus significantly higher than among the normal population.

The dental anomalies are most prevalent in mongols and the association with lenticular defects is also most pronounced in this group.

Idiopathic epileptics and the unclassified mental deficient patients show dental changes in some way different from those encountered in the mongols, but also often co-existing with lens defects.

The mental and lenticular disorders are pre-natal in origin.

The data provide evidence for the fact that the anomalies of the dentition are aetiologically related with the other congenital defects.

It is acknowledged that a defection in the layer or an interference with its normal development will induce concomitant changes in various organs of the same derivation.

Hence, the involvement of the teeth in this process in association with other congenital lesions tends to confirm the argument that the dental changes are part of a syndrome of a systemic condition.

It is considered that the aetiological factors for these disorders are in the ectoderm, and that its inherent characteristics and its mode of development play an important part in the pathogenesis.

Acknowledgment.

It gives us great pleasure to record our indebtedness to Dr. R. M. Stewart, late Medical Superintendent, Dr. J. H. Watkin, Physician-Superintendent of Leavesden Hospital, and Dr. L. T. Hilliard, Physician-Superintendent of Fountain Hospital, who allowed us access to the patients and their records, and who provided every possible facility for the investigation. The medical staff of these hospitals were most generous in their co-operation, and particular thanks are due to Dr. W. M. McGrath.

Above all, we wish to acknowledge our gratitude to Sir Allen Daley, without whose help and support this investigation could never have been made. One of us (R. S.) also wishes to express his very warm thanks to Professor L. S. Penrose and Drs. John Penfold and Herbert Levy for their most valuable advice and interest.

The photographs have been prepared by Miss M. W. Hill and Nucleus Ltd.

References.

Andrews, George Clinton (1946), *Diseases of the Skin.* Philadelphia and London.
Bahr, G. von (1936), *Acta Ophthalmologica,* Supp. **40**, Copenhagen.
Bauer, W. H. (1944), *Am. J. Path.*, **20**, No. 2.
Bellows, John G. (1944), *Cataract and Anomalies of the Lens.* London.
Benda, C. E. (1947), *Mongolism and Cretinism.* London.
Idem (1946), "Ten Years' Research in Mental Deficiency," *Amer. J. Ment. Def.*, **51**, No. 2.
Boyd, W. (1943), *Textbook of Pathology.* London.
Brousseau, Kate (1928), *Mongolism.* London.
Bruckner, M. (1943), "Studies in the Incidence and Cause of Dental Defects in Children," *J. Dent. Res.*, **22**.
Bruno, F. E., and Engelhardt, H. (1944), *Ann. Int. Med.*, **20**, No. 1.
Burks, Barbara S. (1938), *Proc. Nat. Academy of Sciences of U.S.A.*, **24**.
Cockayne, E. A. (1933), *Inherited Abnormalities of the Skin and its Appendages.* London.
Evans. M. W. (1944), "Dental Defects in Infants after Maternal Rubella," *M. J. Australia*, ii, No. 10; (1947), i. 28 June).
Fish, Wilfred E. (1948), *Surgical Pathology of the Mouth.* London.
Gates, R. R. (1946), *Human Genetics.* New York.
Goldman, H. M., and Bloom, J. (1949), *Oral Surgery, Oral Medicine and Oral Pathology*, *St. Louis*, **2**.
Gregg, A. S. (1941), *Trans. Ophth. Soc. Aust.*, **3**.
Gruneberg, Hans (1947), *Animal Genetics and Medicine.* London.
Halperin, S. L. (1946), "Human Heredity and Mental Deficiency," *Amer. J. Ment. Def.*, **51**, No. 2.
Idem and Curtis, G. M. (1942), "Anhydrotic Ectodermal Dysplasia Associated with Mental Deficiency," *ibid.*, **46**.
Ingalls, Th. H., and Gordon, J. E. (1947), "Epidemologic Implications of Developmental Arrests," *Am. J. Med. Sci.*, **214**.
Keith, Sir Arthur (1948), *Human Embryology and Morphology.* London.
Kreyenberg, In: Bumke, O., and Foerster, O. (1936), *Handbuch der Neurologie*, **16**.
Lennox, W. G. (1947), "The Genetics of Epilepsy," *Am. J. Psych.*, **103**.
Idem (1947), "Epilepsy," *Proc. Ass. Res. Nerv. and Ment. Dis., Baltimore.*
Idem and Gibbs, E. (1944), *Trans. Amer. Neurol. Assoc.*

LOWE, R. F. (1949), " The Eye in Mongolism," *B. J. Ophth.*, **33**, No. 3.
MANN, I. (1937), *Developmental Abnormalities of the Eye*. Cambridge University Press.
MELLANBY, H. (1941), " Effect of Maternal Dietary Deficiency of Vitamin A on Dental Tissues in Rats," *J. Dent. Res.*, **20**.
MELLANBY, (Lady) M. (1934), " Diet and the Teeth," *Med. Res. Council, Spec. Rep.* No. 191, Part 3.
MOORE, ROBERT A. (1944), *Textbook of Pathology*. London.
PARSONS, Sir J. H., and STALLARD, H. B. (1942), *Diseases of the Eye*. London.
PENROSE, L. S. (1934), *The Influence of Heredity on Disease*. London.
SARNAT, B. G., and SCHOUR, J. (1941), " Roentgenographic Diagnosis of Congenital Syphilis in Unerupted Permanent Teeth, "*J.A.M.A.*, **116**.
Iidem (1941), " Enamel Hypoplasia in Relation to Systemic Disease," *J. Am. Dent. Ass.*, **28**, 1941, and **29**, 1942.
SCHEER, W. M. VAN DER (1927), *Abh. Neurol. Psychiatr. Psychol.*, **41**.
SEQUEIRA, J. H., INGRAM, J. T., and BRAIN, R. T. (1947), *Diseases of the Skin*. London.
SNYDER, L. H. (1941), *Medical Genetics*. Duke University Press.
SPITZER, RICHARD (1933), " Enamel Hypoplasia in Idiopathic Epilepsy," *Deutsch. Med. Woch.*, **59**, and *Brit. Med. J.*, i, 1942.
STONES, H. H. (1948), *Oral and Dental Diseases*.
SWAN, CH., TOSTEVIN, A., *et al.* (1943), " Congenital Defects in Infants following Infectious Diseases during Pregnancy," *Med. J. Austr.*, ii, No. 11, and *ibid.*, ii, 1946.
THOMAS, D. H. H. (1939), " Anodontia in Mongolism," *J. Ment. Sci.*, **85**.
TREDGOLD, A. F. (1947), *A Textbook of Mental Deficiency (Amentia)*. London.