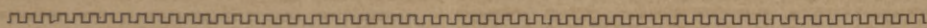


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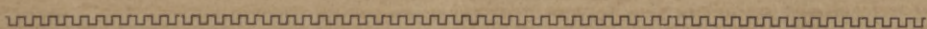
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FOUND IN THE SO-CALLED MONGOLIAN
TYPE OF IDIOCY.

BY CHARLES A. OLIVER, M.D.,
Attending Surgeon to Wills' Eye Hospital, Philadelphia, etc.



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A CLINICAL STUDY OF THE OCULAR SYMPTOMS FOUND IN THE SO-CALLED MONGOLIAN TYPE OF IDIOCY.¹

BY CHARLES A. OLIVER, M.D.,

Attending Surgeon to Wills' Eye Hospital, Philadelphia, etc.

THIS paper, which is the outcome of clinical work² extending over a period of nearly three years at the Pennsylvania Institution for Feeble-minded Children, at Elwyn, Pa., and which embraces, to a greater or a less degree, many of the peculiar ocular groupings found in this so-called class of cases, is here given in the hope of not only adding a few of the more easily seen ophthalmic symptoms to the main characteristics of the disorder, but that some additional proof may be furnished as to the true pathology. Reserving the details of the latter examination for another paper, the subject will here be confined to a series of short clinical observations in association with a definite number of deductions upon the eye conditions alone.

Although classed by most alienists and neurologists as a genetous form of idiocy, in which the physiognomy is distinctly Mongolian in type—a form of ethnic naming, however, which, with equal force, might be given to those cases presenting other racial facial characteristics—yet, in this brief clinical demonstration of some of the ocular groupings found, the ophthalmic conditions have been so constant, and are so different from those that are seen in the mentally and physically healthy of our race, that they deserve to be specially studied from this standpoint. The paper, therefore, resolves itself into a study of the eye symptoms of that peculiar form of congenital idiocy which presents a marked brachycephalic deformity as one of its most prominent features.

Governed by the same rules of selection of cases as in all of the previous work in this direction by the writer,³ and care having been taken that all of the conditions were studied under similar circumstances, the observations have been combined into fifty-seven short captions, from which five conclusions will be formulated.

OBSERVATIONS.

First.—Direct vision for form in the couple of cases in which it could be studied, slightly below normal.

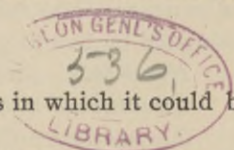
Second.—Direct vision for color slightly subnormal in one instance in which it could be properly obtained.

Third.—Accommodative action: impossible to obtain any reliable answers by any subjective or objective methods.

¹ Paper read before the American Ophthalmological Society, September 24th, 1891, at the Second Triennial Meeting of the Congress of American Physicians and Surgeons.

² The writer desires to express his thanks to Drs. I. N. Kerlin and A. W. Wilmarth for their kindness in allowing him access to the proper class of subjects, and to Drs. J. S. Stewart, L. F. Love, H. W. Cattell, F. P. Norbury and M. W. Barr, for assistance given.

³ *Vide* Transactions of the American Ophthalmological Society for 1887, 1889, and 1890.



Fourth.—Visual fields: nothing could be properly gotten.

Fifth.—Circumference of orbital cavities probably somewhat larger than normal, the bony rim in many cases being unusually thin.

Sixth.—Bony bases of orbital cavities somewhat squared, asymmetrical, and situated farther apart than normal.

Seventh.—Greatest vertical diameter of the circumference or base of the bony orbit in a number of cases, directed slightly down and out, and away from the median line.

Eighth.—Greatest lateral diameter of the circumference or base of the bony orbit, in a number of cases, directed slightly in and down, and toward the median line.

Ninth.—Superciliary margin of orbit slanting upward and outward, causing, in many instances, a distinct lessening of the usual downward inclination of the outer third of the supercilia.

Tenth.—Palpebral fissures, in every instance, obliquely placed toward one another, deviating downward and inward toward the median line, and at angles varying from five to twenty degrees with the horizontal meridian.

Eleventh.—Palpebral fissures generally placed at different angles in relation to one another, that of the right eye usually showing the greater inclination.

Twelfth.—Palpebral fissures, as a rule, corresponding in direction with that of the greatest lateral diameter of the orbital base.

Thirteenth.—Palpebral fissures, in every instance, quite short, averaging but twenty-three to twenty-four millimetres in length.

Fourteenth.—Palpebral fissures, in a number of cases, differing about a millimetre in length in the same subject.¹

Fifteenth.—Horizontal plane at the position of the right inner canthus, somewhat more frequently lower than that at the left inner canthus.

Sixteenth.—Lid-substance, in a few instances, tumefied and soft to the touch; the inferior orbital and the orbito-malar sulci being not very clearly defined.

Seventeenth.—Ciliary borders of lids thickened and broad in quite a number of cases.

Eighteenth.—Conjunctivæ very vascular, swollen and excreting; the orbital portions of the membrane being very frequently succulent—these conditions increased by handling.

Nineteenth.—Follicles in the lower cul-de-sacs very much enlarged.

Twentieth.—Plica semilunaris thicker than ordinary in several instances.

Twenty-first.—Epithelial layer of cornea thickened, filled with rugæ, and, in some instances, dotted with minute blebs.

Twenty-second.—Horizontal diameter of corneæ, in a number of cases, at different angles from that of the palpebral fissures.

Twenty-third.—Pupillary openings three millimetres in average size.

Twenty-fourth.—Interpupillary distance, as a rule, about fifty millimetres in length.

¹ It is curious, though it may be a coincidence, that the left palpebral fissure was almost always the longer.

Twenty-fifth.—Irides, as a rule, freely and equally responsive to light-stimulus.

Twenty-sixth.—Irides, in the few instances in which the experiments could be tried, not very prompt to efforts for accommodation and convergence.

Twenty-seventh.—Irides utterly devoid of any peculiarity in comparative tint.

Twenty-eighth.—Pupils dilated almost *ad maximum* by the instillation of three drops of an eight-grain solution of hydrobromate of homatropine.

Twenty-ninth.—Extraocular motion intact in all directions in the two instances in which it could be properly studied.

Thirtieth.—Slight insufficiency of the interni manifest in the one case (a low grade, with six diopters of hypermetropia) in which it could be satisfactorily searched for.

Thirty-first.—Horizontal nystagmus, ataxic in variety, in several cases in which the deeper temporal layers of the optic nerve tissue appeared very gray and degenerate, and the choroid and retina much disturbed.¹

Thirty-second.—Intraocular tension normal.

Thirty-third.—Intraocular media, in all instances, clear and transparent.

*Thirty-fourth.*²—Optic disks, in a number of cases, unequally grayed, especially in the deeper layers and to the temporal sides; this being more marked where the outlying choroid and retina seemed the most disturbed.

Thirty-fifth.—Substance of disk, in the great majority of instances, especially where the retinal and the choroidal disturbance was not pronounced, apparently oedematous and superficially overcapillary.

Thirty-sixth.—Edges of disk, except to the temporal side, almost completely hidden at first view, but dimly seen by careful and concentrated focusing upon the successive parts.

Thirty-seventh.—Scleral ring quite narrow, and as a rule, only seen to the outer side of the disk.

Thirty-eighth.—Crescents and broken areas of blackish pigment, generally limited to the temporal edge of disk.

Thirty-ninth.—Physiological excavation in most instances seemingly filled in with an almost transparent yellowish gelatinous-like mass.

Fortieth.—Retinal striation but very slightly marked, it always being more pronounced just beyond the superior and the inferior borders of the disk.

Forty-first.—Retina surrounding the optic nerve head oedematous and swollen in quite a number of cases.

Forty-second.—Retinal vessel walls, where they could be seen, especially on the disk, extremely thin and apparently somewhat yellowish and translucent, appearing as if they had been soaked.

Forty-third.—Retinal blood-currents excessively pallid.

¹ This nystagmic action was, in three instances, best noticed during ophthalmoscopic examination made through an artificially dilated pupil. It could be momentarily quickened by directing the attention of the subject to some brilliant object.

² All of the intraocular structures below described were most carefully studied after enlarging the pupillary area, thus giving a broad and extensive ophthalmoscopic field.

Forty-fourth.—Light-streak along the retinal vessels, in a number of cases, quite broad, uniform and clearly defined, whilst in some others, especially in those where there were many evidences of inflammatory or exudative products, it was probably narrower, more irregular, and less visible.

Forty-fifth.—Retinal veins, in the majority of cases, tortuous in appearance.

Forty-sixth.—Macular twigs of the retinal circulation, in a few cases, apparently larger than ordinary and more plainly visible than usual.

Forty-seventh.—A fine network of minute branching vessels having no apparent communication with any other retinal vessel grouping in the macular region, in two cases where the retinal swelling was quite dense and extensive.

Forty-eighth.—“Shot-silk” opacities, so ordinarily seen in children, not very pronounced and not numerous.

Forty-ninth.—Small clusters of pin-point spots apparently like the remains of minute capillary hemorrhages, seated superficially in the retina in many instances, these more generally following the course of some of the smaller inferior retinal stems.

Fiftieth.—Isolated and aggregated cholesterin crystals in some instances, generally situated in positions bearing no relation to the larger retinal vessels.

Fifty-first.—Whitish and yellowish massings, from the size of pins' heads to two or even three millimetres in area, irregularly distributed throughout the retina in a number of cases.

Fifty-second.—Rim of macula lutea itself quite large, irregular and apparently stretched laterally in a few cases where there was not much intraocular disturbance, especially where there were low degrees of hypermetropic and myopic refraction, with and without noticeable astigmatism.

Fifty-third.—Border of yellow-spot, in a few instances densely white in appearance.

Fifty-fourth.—Pigment layer of the retina reduced in thickness and density, allowing the larger choroidal vessels to be plainly seen in the periphery of the eyeground.

Fifty-fifth.—Choroid, in a great number of cases, disturbed, granular, and irritated.

Fifty-sixth.—Choroid, in some instances, seemingly in a low-grade, inflammatory state, the greatest amount of disturbance being in the macular region and vicinity of the disk.

Fifty-seventh.—Refraction in the macular region, generally hypermetropic, with some astigmatism.

In more than fifty per cent. of the cases examined in this grouping, post-mortem examination showed a decided lessening in both numbers and size of the ganglion cells in the hemispheres, especially in the frontal regions, widely scattered sequelæ of cerebral vasculitis, and marked deficiency of the pons and medulla.¹ These are of extreme interest in exhibiting the more pronounced conditions of the intracranial contents, which may be either parts and parcels of the general systemic involvement, or merely anatomical and pathological

¹ *Vide* Wilmarth, pages 59 and 60, Proceedings of the Association of Medical Officers of American Institutions for Idiots and Feeble-minded Persons, 1887 and 1888.

peculiarities due to defective development of the nutritive centres in the lower brain.

Here, where, gathered from every phase of social condition, we have a definite and an easily recognized class of subjects, dwarfed, stunted, and peculiarly shaped, markedly brachycephalic or even plagiocephalic, and universally almond-eyed¹—a type of patients with cold, club-shaped extremities that must be wrapped during the moderate winter of our latitude, to prevent them from being frost-bitten—it must be conceded that there is a something which is peculiarly common in their physical formation. Moreover, here where there is a grouping of cases in whom mere scratches become ulcerous, sores fail to heal until warm weather appears, and ecchymoses are so prevalent during sickness—a type of patients who, without any history of ancestral ties of consanguinity in marriage, almost always succumb during one of the colder months of the year to some gross hemorrhagic or exudative lesion in the mucous tract—it must be agreed that there is some particular form of dyscrasia which is alike in all; and if this class of individuals present the above ocular conditions, so pronounced, so indicative of the general symptoms, and so unlike those seen in any other form of idiocy, it can be justly claimed that in them there has been some common etiological factor which has been quite actively at work during the very early history of their antenatal existence.

So regarding these cases, then, the following deductions may, with some show of certainty, be tentatively made—conclusions that, if at all certain and sure, assume an interest not only to the ophthalmologist, but a direct value to the alienist, the neurologist, and the pathologist—a series of results, correct or incorrect, that at least may be of service in the inducement for further study in this and other directions of the many types of cases which apparently present similar groupings of coarse and, at most times, easily recognized peripheral lesions.

CONCLUSIONS.

I. In the so-called Mongolian type of idiocy, the malpositions, irregularity of contour, and inequality in comparative size of the bony orbits, with the obliquity of the attached ligamental and tarsal tissues, giving the palpebral fissures their peculiarity of direction, the lids their shortness, and the eyes their apparent relative faulty situations, are merely the rough ocular expressions of the results of the osseous and ligamental malformations so characteristic of the disorder: in fact, it is these conditions which have, more than any other, contributed to the naming of the type.

II. In the so-called Mongolian type of idiocy, the ocular bulb, in nearly every instance, presents peculiarities of structural change characterized by the appearances of the results of low and chronic forms of neuro-retinitis and choroiditis, indicative of local inflammation of these parts, both before and after birth of the individual.

III. In the so-called Mongolian type of idiocy, the substance of the intra-ocular ending of the optic nerve and the circumjacent retinal and choroidal

¹ Not strictly so, as the fissures are short.

membranes, seem, in those instances where there are no marked evidences of coarse intraocular change, to share in the soft, jelly-like œdema so universally recognized in the external portions of the organ.

IV. In the so-called Mongolian type of idiocy, the mucous lining of the ocular appendages and of the anterior part of the globe, in every instance, exhibits the many gross pathological peculiarities seen in the chronic and constantly provoked inflammation of other similar surfaces found throughout the same subject, and which in the great majority of cases leads to lethal result.

V. In the so-called Mongolian type of idiocy, the peculiar vascular changes, not only discoverable—even ophthalmoscopically—in the vessels of the retina, where the visible portions of the walls appear thin and so peculiarly tinted, and the sequelæ of fine capillary and even larger hemorrhagic extravasation into the retinal substance, with probable evidences of graver complications, are most common, but, as almost universally seen in identical imperfections and disease of intracranial vessel structure,¹ serve, with the general clinical features of imperfect circulation, to show, both objectively and subjectively, the prominent characteristic lesion, and probably etiological condition of the disorder, *imperfect development with consequent disease of the entire vascular system.*

¹ For comparison with the faulty conditions of the retinal structures, a careful comparison of the related cortex cells in the occipital region would be most interesting and instructive: in fact, all of the structures, both neural and vascular, can be studied to advantage.

It may be of interest to note that the average age was twelve years; but one case gave any evidence of organic cardiac disease, though the heart, in several instances, examined post mortem, was found to be quite small; in all but two cases—both of whom were paralytics—the patellar-tendon reflexes were either absent or greatly diminished; and, as a rule, the grandparents, more particularly upon the maternal side, gave a history of paralysis.

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