Kalmuc idiocy: report of a case with autopsy / by John Fraser; with notes on sixty-two cases by Arthur Mitchell.

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17

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REPORT OF A CASE WITH AUTOPSY.

By JOHN FRASER, M.B. -

WITH NOTES ON SIXTY-TWO CASES.

By DR. ARTHUR MITCHELL,

Commissioner in Lunacy.

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KALMUC IDIOCY: REPORT OF A CASE WITH AUTOPSY.

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Commissioner in Lunacy.

My attention was directed to this case by Dr. Arthur Mitchell, Commissioner in Lunacy, in the course of his official visit last spring. He informed me that it was one of so-called Kalmuc idiocy, a form of idiocy rarely met with in asylums, but nevertheless not really uncommon. Mitchell pointed out in this case the distinguishing physical characters of this class of idiots. Some of the more prominent of these are obliquity of the eyes and palpebral slits, shortness and flatness of the nose, depression at its root, broadness or squatness of the hands, transverse fissuring of the tongue, shortness of stature, disproportion between different parts of the limbs, smallness of the head, and shortness of its antero-posterior measurements. Certain mental characteristics are said to be very constantly present, and these will be referred to in the details of the case. The peculiarity of the eyes and nose give the face a Kalmuc look, and hence the name.

After having my attention thus drawn to this case, I endeavoured to obtain the literature relating to this form of idiocy, but without success. Dr. Mitchell, however, has notes, casts, photographs, sketches, &c., of a considerable number of cases. He has given me in a letter a digest of these notes which he has permitted me to append to this report. The lithograph is from a photograph kindly lent me

by Dr. Mitchell.

Report of Case.

Elizabeth Meldrum, aged 40; admitted 18th Feby., 1875; from Ferryport-on-Craig, Fife; died, 29th March, 1875.

History.—The following is all I have been able to gather regarding the patient. She was an illegitimate child, and parturition at her birth is said to have been normal. Her mother, who died some time before the patient's admission into the asylum, is said to have suffered remorse and mental pain during her pregnancy, and to have expiated her sin by constant and affectionate care of her unfortunate offspring.

Physical Condition.—Patient was a short slenderly made woman in fair condition, weight 6st. 12lbs., height 4ft. 81 inches. trunk was symmetrical but flattish; breasts small and firm, resembling those common in a girl in 12 or 14; upper limbs were short; arm very short in comparison with forearm; hands broad and short: fingers short, and when hand was placed on any flat surface, the fingers inclined to spread widely out; joints of fingers lax, and admitted of abnormal extension, nails almond-shaped, short and curved inwards at points. Lower limbs were short in proportion to the trunk; thigh short in proportion to the leg. The foot presented the following peculiarities: the big toe was large in proportion to the foot; second toe was unusually long, third toe was half the length of the second the fourth, half the length of the third, and the fifth or small toe was of normal size (see sketch). The fourth toe was curved downwards. and but a small triangular portion was left uncovered by the third and small toes. This peculiar deformity was symmetrical, and from the drawings of the feet of other Kalmuc idiots, it seems a deformity not altogether uncommon among them.

Her head was small and round. The asymmetrical condition of the head, which will be particularly referred to in the description of the skull, was not observed during life. The hair was fine, thin in quantity, and of a light brown colour. The ears were small and

simple in conformation.

The following are the dimensions of the head :-

1.	Circumference in line of upper part of ear and	eyeb	rows, ir	iside	Inches.		
	berance .	. 0001	Prour Pr	ota	191		
2.	From root of nose to occip	ital r	rotuber	ance	113		
3.	From ear to ear over vault				101		
4.	Longitudinal diameter		- 70	-	$6\frac{7}{4}$		
5.	Transverse .				64 55		
6.	Diagonal diameters—						
	From right anteriorly to left posteriorly						
	,, left anteriorly to	right	t poster	iorly	578		

Dr. Mitchell pointed out to me that the plane of the face and the plane of the back of the head tended to form two parallel lines. The space between these lines measured 63 inches.

The face was almost square, the length to the breadth being 6 inches to 43. Cheek bones prominent, and chin sharp and projecting Forehead was rounded in outline and surface, and rose in a moderately

straight manner. It was by no means a retreating or low forehead. Two peculiarities were at once noticeable, viz., the obliquity of the palpebral slits and the broadness of the base of the nose. The palpebral slits ran outwards and upwards in a very marked manner. The inner canthus seemed drawn down. The eyebrows shared in this obliquity. The eyes were small in comparison with the size of the orbits, colour of iris grey; and eye-lashes were absent. The bridge of the nose was narrow, but inferiorly the base became broadened out. Mouth was small, and always pursed, lips thin and angles generally curved downwards.

Interior of mouth presented the following peculiarities: the tongue was long, thin, and much pointed. On the dorsum were numerous transverse fissures, and on the left side about the beginning of the posterior half was a strip of smooth surface about an inch long, and three-eighths of an inch broad. Only the incisor teeth were present, two in upper and four in lower jaw. Palate was not particularly arched.

Physical examination as to the position, size, and condition of in-

ternal organs revealed nothing abnormal until her illness set in.

Mental Phenomena.—Her intellect may be said to have been that of a child from a year to eighteen months old. She could not speak, but she uttered sounds with volubility as if she were busily speaking, and when in anger she did this with emphasis and vehemence. Her sight and hearing were good. She remembered the faces of those who were kind to her, and of those who annoyed her, and sought notice from the former and avoided the latter. Her chief characteristic was an affectionate disposition. This was evidenced by her kind, contented, and happy expression, and by her grasping the hand of any one who took notice of her, patting it, and putting it to her cheek. At times she had the peculiar habit of putting one's hand on the back of her head, and indicating that she wanted it smoothed.

Another characteristic was her love of decoration. Any bright article of dress she wore with jealous care, and drew every one's attention to it. If any other patient had anything gay on, she always

pointed to it.

She is reported as being very fond of music.

She continually sat in the corner of a bench next the fire, with her feet under her. She had no sense of modesty, and her habits were dirty. She had a great hatred of water, and her struggles against being bathed were strong and persistent. On admission she was extremely dirty, and I attribute the cause of death, acute pleurisy, to the constant bathing which was rendered necessary by her habits.

The description of the mental state would have been fuller, had the patient been longer under observation. She died after about six

weeks' residence in the asylum.

Post-mortem Examination.—After the full details given under the head of "physical condition," I need here only describe the skull and brain, and incidentally refer to the viscera within the trunk.

Head .- Scalp very thin. (Sketch of cast of head as taken 14)

hours after death is to be seen in Plate 2.)

Skull as a whole is very light, weighing, inferior maxilla included, only 1lb. Many parts are extremely thin, especially the roofs of the orbits, the wings of the sphenoid, and the fossa for the lobes of the cerebellum.

The configuration is asymmetrical. The cranium seems as if it had been grasped diagonally from left anteriorly to right posteriorly, and squeezed. There is a flattening over the outer edge of the left orbit and right parietal eminence, and a slight bulging out at right frontal region and left parietal eminence. The internal fossa are unequal, the left occipital fossa is $\frac{3}{8}$ of an inch broader in the transverse direction than the right, and the right middle fossa is evidently broader than the left.

The sutures of the vault are very open; slight force would separate each bone. The frontal is open throughout its entire length, though the age of the patient was forty. As many as five wormian bones are to be counted in each lambdoidal and squamous suture. At the base, a wide interval exists between the petrous bones and the basilar portions of the occipitals. In these spaces, wormian bones of apparently poor cancellous tissue are to be found. The spheno-basilar synchondrosis is not visible, nor is there any elevation or ridge to mark its site. The basilar bone seems normal in every particular. The suture between the lesser wings of the sphenoid and the frontal bone is open. From the state of all the sutures, it will be evident that the skull could have expanded in every direction. This case is therefore one not due to early ossification of any suture.

Foramina of the base are large, especially the median and posterior

on either side. Arterial foramina are normal.

As regards individual bones, the most important peculiarity is the absence of the nasal bones. A small projection from the frontal bone seems to be their representation. (Well illustrated in sketch of skull.) The processus auditorius on each side is whiter than the surrounding parts of the temporal bone, and these processes seem to have maintained their individuality. The horizontal plate of the palate bone is not arched, and the suture between the two maxillary bones at this part is complete. The inferior maxilla has the obtuse angle consequent upon an edentulous condition.

The orbital cavities are normal, but the anterior nasal aperture is,

comparatively speaking, large.

The following are the measurements of skull :-

Measurements of Skull.

_			Inche			
External	cranial	circumference	e			171
"	"	longitudinal	curve			1118
"	,,	transverse	,,			111
Longitu	dinal dia	ameter ·		1.		57

				Inches.
Transverse diameter				$5\frac{1}{4}$
Bimastoid "				4
Bizygomatic ,,		36		45
Frontal "				$3\frac{15}{16}$
Length of Hard Palate			**	45
Breadth ,, at	t widest p	art.		14
Breadth of Orbit .				176
Vertical width of Orbit				176
Depth of Orbit.				1_{16}^{9} $\frac{78}{8}$ 1_{16}^{7}
Width of Anterior Nares		t part		8
Vertical width of Anterio	or Nares			176
Width between orbits				34
Internal Base—	~			
Distance from Foramen		to Inte	rnal	-,
Occipital Protuberar				$5\frac{1}{4}$
Distance from Foramen		to Ant	erior	0.1
edge of Foramen Ma		. 10		316
Distance from junction of			inus	4.1
to Lateral Sinus on				41/4
Anterior-posterior length				11/4
	ter of Fo		mag-	1.5
		Vacuum		15
Transverse diameter of I	oramen 1	Magnun		1 5

Brain.

The membranes were normal. Veins running into Sup. Longitudinal Sinus were engorged in consequence of the obstructive pulmonary disease of which the patient died.

The organ is small, the whole encephalon only weighing $40\frac{1}{8}$ ozs. Consistence was normal, but configuration was asymmetrical, corresponding, of course, to want of symmetry described as appertaining to the skull. There was bulging in right frontal and left parietal regions, and slight flattening in left frontal and right parietal lobes.

Convolutions were slightly flattened when brain was removed. The general course of the gyri is comparatively simple. (See drawing of brain.) The sulci vary in depth; anteriorly many are very deep. The principal fissures are well marked, the external parieto-occipital fissure on the left side runs outwardly for nearly two inches. Posterior part of both third frontal convolutions is very narrow; this taken in connection with the patient's want of articulate speech is interesting. The island of Reil on both sides is small and undivided; no convolutions spring from its external margin. The superior gyrus of both temporo-sphenoidal lobes is very narrow.

The base was carefully examined at time of removal, but the cranial nerves, pons, medulla, crura, and arteries presented no noticeable

abnormality.

On Section, the grey matter had a breadth of four to five millimetres anteriorly, and of three to four posteriorly. Being desirous of preserving the brain in its entirety, only few measurements with the tephrylometer have been taken, and only the tip of the right frontal lobe was hardened for sections.

Ventricles normal.

Microscopic Examination of grey matter shows an average number of tripolar and multipolar cells of apparently normal size, distinctness, and contents. Some have the usual yellow pigment. Nuclei of neuroglia are not increased, nor is there any abnormal fibrillation. The perivascular canals are dilated, and the hyaline membrane is in many preparations puckered and kinked.

Cerebellum.—The left lobe is slightly longer in transverse diameter

than the right, otherwise this organ is normal.

Sympathetic ganglia of the neck were removed. These were small, and the right superior one was half the size of the one on the left side.

Trunk.

Thorax was symmetrical.

On opening cavity, 75 ounces of bloody serum was found in left pleural cavity. Both costal and visceral pleuræ were coated with vascular flocculent lymph; organ collapsed. On section, substance was found to contain miliary tubercles in first or cartilaginous stage.

Right lung was emphysematous anteriorly, and hyperæmic pos-

teriorly. Miliary tubercles also present, but in lesser quantity.

Heart was small, aortic valves fenestrated, otherwise normal.

Abdomen.

Though every organ was carefully examined, it is unnecessary to detail the state of each. I may mention, however, that the uterus was small, or round, cervix uteri constricted, and that the ovaries were small, and full of cicatrices of ovulation, but none apparently recent.

Notes by Dr. MITCHELL.

1. I have a record of visits paid to sixty-two so-called Kalmuc idiots; and I have notes, more or less full, regarding the physical and mental condition of fifty-four of the sixty-two. What follows is taken from these notes, and refers of course to the class rather than to individuals.

2. I think it almost beyond question that they are more

common on the east side of Scotland than on the west.

3. I have never seen a Kalmuc idiot in any Scotch asylum, except in that of the Fife and Kinross District, in which there have been two—Kinloch and Meldrum. In our insti-

tutions for imbeciles, so far as I can remember, I have only seen two.

4. Of the fifty-four, thirty were males and twenty-four females.

5. I think it nearly certain that they are short-lived. The oldest I ever saw was 43. The ages of the fifty-four are shown in the statement which follows:—

AGES.		M.	F.
0-10	 	 7	0
10—15	 	 5	6
15-20	 	 5	4
20-25	 	 6	4
25—30	 	 0	-4
30—35	 	 5	4
35—40	 	 1	1
40—45	 	 1	1
		-	_
		30	24

6. Many of the fifty-four are now dead. Phthisis caused a large majority of these deaths. In not a few instances, however, death was attributed to nothing more definite than general decay—a sort of precipitated senility.

7. I have recorded the height of seven adult males and fifteen adult females. The males give an average of nearly 4 feet 8 inches—the range being from 4 feet 2 inches to 5 feet 3½ inches. The females give an average of nearly 4 feet 5 inches—the range being from 4 feet 1 inch to 5 feet 1 inch.

8. I have various head measurements in the cases of eleven adult males and fourteen adult females. The average circumference of the male heads was $20\frac{3}{8}$ inches, and the range was from 19 to $21\frac{1}{2}$. The average circumference of the female heads was $19\frac{4}{8}$, and the range from 18 to $20\frac{1}{2}$. The antero-posterior relatively to the lateral diameters were short—that is, the heads were brachycephalic, as well as small. I think this was almost invariably the case.

9. In a large proportion of the cases, these idiots were recognised as notably small and weak at birth. I convinced myself that this was almost certainly true of nineteen of the males and thirteen of the females. In other words, the condition appears to date from the intra-uterine life.

10. Heredity as a cause does not appear with any special frequency in their history. My notes rather indicate the reverse.

11. Kinship of parentage is certainly infrequent. No case of the relation of full cousinship between the parents is recorded in my notes. In four cases, however, the parents were related to each other as second cousins, and in other two cases a still more distant relationship existed. On this point I obtained information regarding fifty-one of the idiots.

12. Of a considerable number I have noted that the mothers were in a marked state of bad health during pregnancy; in a state of bad health, however, which received no name, but of which it was always told to me that emacia-

tion was a result.

13. It not unfrequently happens that in the male adult Kalmuc idiot only one testicle descends. I have recorded this of five cases. In one boy, ten years old, neither testicle had come down. The hair on the pubes, in the axillæ, and on

the chin and cheeks, is generally scanty.

14. Puberty is reached late and the menstrual discharge soon ceases in the females, who never show active eroticism. My observations point clearly to this conclusion as correct. I have not known any case of a Kalmuc idiot woman having a a child. As in the males, so in the females also, the hair on the pubes and in the axillæ is scanty. At the time of the establishment of the menstrual discharge, however, plumpness appears, and the mammæ are fairly developed.

15. The shape of the head—the position of the eyes—the weak sight—the condition of the tongue, teeth, and palate—the burr in articulation and the defective speech—the shape of the hand snd feet—&c., &c., occur in the class generally very much as they occurred in Meldrum, but they are better seen in idiots of the class who are somewhat younger than she was, and who have not begun to show signs of decay.

16. The large, rough, red, hacked tongue is very constant. At one time I thought there was a uniformity of pattern in the hackings, but the drawings I made show that this is not

the case.

17. In no instance did I find that the hand or foot differed in character from what was seen in the case of Meldrum. As I made numerous casts and drawings I was led to look minutely into this point. I refer here only to the shortness and broadness of the hand and foot.

18. Many of the fifty-four were photographed for me, and of others I had large chalk drawings made. Both the photographs and drawings were the work of country artists, and with the exception, perhaps, of the little photograph I

gave you (the name of patient I have omitted to preserve) not one of them can be called successful.

19. The concurrent constancy of a characteristic condition of mind with a characteristic condition of the body is what gives interest to this form of idiocy. The mental state is as distinct, as peculiar, and as steady as the physical. If the fifty-four were brought together they would be found to resemble each other strikingly in personal appearance. But more than this, they would also be found to resemble each other in character, in capacity, in likings and dislikings, in

habits, in defects, in aptitudes, &c.

20. They are not cretins, but I can quite understand that they might be held to be cretinoid idiots. Of full cretins there are, I think, only from 12 to 20 in Scotland, appearing preferentially in no particular district. Not one of the full cretins seen by me in Scotland was goitrous. The same was true of the Kalmuc idiots. But both full cretins and cretinoid idiots occur, even in strongly goitrous districts, without exhibiting in themselves any trace of goître. Cretinism is a form of idiocy which affects goitrous districts, but which may present itself anywhere. When goître is associated with cretinism, it is as an accident of the idiocy. That accident, of course, will present itself with frequency where goître prevails, which it usually does where cretinism pre-Goitrous persons are not necessarily cretins or cretinoid; and cretins or the cretinoid are not necessarily goitrous. It is even a question whether in a goitrous district, the proportion of the cretins who are goitrous is very much greater than the proportion of the goitrous in the general community. It is quite possible, therefore, to regard the Kalmuc idiots as cretinoid, though they show no tendency to goître. I do not say that they should be so regarded; but at the same time I recognise certain points of alliance between the mental and physical states of so-called Kalmuc and of cretinoid idiots.

21. Only two of the fifty-four are recorded as illegitimates, but I scarcely think that this can represent the case truly. Of idiots and imbeciles of all sorts in Scotland, one in every six is born out of wedlock. There can, however, be no special connection between illegitimacy and Kalmuc idiocy, for (putting aside the fact that I have not recorded anything to show such a connection, which might be an omission, and is at best negative evidence), I find that I have in a large number of cases recorded the idiot's position in the family

of which he was a member—that is, whether he was first born, last born, or neither first nor last. The facts so recorded do not point to the frequent occurrence of illegitimacy.

22. Some of these facts are grouped in what follows :-

(a) Six of the idiots are given as first born.

(b) Twenty as the absolutely last born in families consisting of more than one and often of many more than one child.

(c) Eight as not the absolutely last born, but as appearing

far down among numerous births. Thus:-

1 is said to be the sixth child
1 ,, ,, seventh child.
4 are said to be ninth children.
1 is said to be the tenth child.
1 ,, , fourteenth child.

My notes indicate that in these eight families other births followed the birth of the idiot,—even in the last, where the idiot was a fourteenth child.

I have not noted anywhere that the idiot was a solitary child. Still in some cases that must have been so, as happened indeed in the case of Meldrum, who is one of the two illegitimates referred to.

The inference from these facts appears to be that Kalmuc idiots show even a less tendency to be first born children than do the idiotic taken all together. Illegitimates are

frequently first born.

- 23. In thirteen instances I have noted that at the birth of the idiot labour was normal and easy. In no case have I noted that instrumental assistance was needed. This I think conclusively shows that Kalmuc idiocy is not referable to injuries sustained at birth, because, when I made these notes, I took a special interest in the whole child-bearing history of the mothers of idiots, including in a special manner the birth-history of the idiots themselves. On these points, I subjoin all the facts which I have recorded, and which seem of any value:—
 - 1. In one case there were long and irregular intervals between the births of the different members of the family.

In another case the idiot was born prematurely.
 In another, the idiot lost blood largely from the navel soon after birth.

4. In another, the idiot presented by the breech, and the labour was long.

5. In another, the mother was seriously injured by a fall

when pregnant with the idiot.

6. In another, the idiot was born eleven months after its mother had had a miscarriage in the third month of pregnancy—the miscarriage being followed by illness and great debility.

7. In another, the idiot was born two years after its mother

had had two miscarriages close after each other.

8. In another, frequent miscarriages appear in the child-

bearing history of the mother.

- 9. In another, the mother, after bearing six healthy children, was injured by a fall—had several miscarriages—then ceased to become pregnant for eight years, when she gave birth to an idiot, and after another short interval to the Kalmuc idiot.
- 10. In another, labour was long, and the placenta adherent.

11. In another, after having several healthy bastards, a woman married, and bore the Kalmuc idiot—the only

child she had in marriage.

12. In another, the parents of the idiots were five years married before the birth of their first child. After another interval of five years, their second and last child, the idiot, was born.

13. In many cases the mothers told me that the motion of

the idiot in utero was little felt.

- 14. In one case, the mother of the idiot ceased to be prolific at 31.
- 15. In another, at 35.

16. In nine cases the mothers' ages at the time of the idiots' birth were respectively 50, 46, 44, 43, 43, 42, 42, 42, 40.

17. In one case the father's age was said to be 70.

- 18. In another, the mother of the idiot had twins, and so had its father's mother.
- 19. In another, the idiot was one of twins.

20. In another, the father's mother had twins.

21. In another the mother of the idiot had twins, and the maternal aunt of the idiot had twins four times running.

22. In another, the mother of the idiot had twins.





