## Cases of sporadic cretinism treated by thyroid extract / by Telford Telford-Smith.

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## **Publication/Creation**

Lewes: South Counties Press, [1895]

## **Persistent URL**

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## CASES OF SPORADIC CRETINISM TREATED BY THYROID EXTRACT.

BY

TELFORD TELFORD-SMITH, M.A., M.D.,

Medical Superintendent, Royal Albert Asylum, Lancaster.

Reprinted from " The Journal of Mental Science," April, 1895.

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PLATE I.



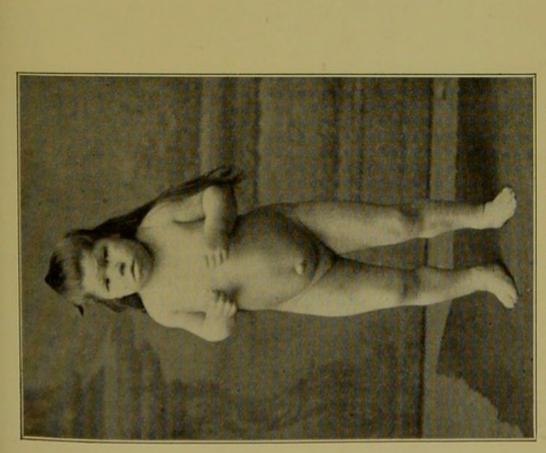
GROUP OF CRETINS (AFTER TREATMENT.)

To illustrate Dr. Telford Smith's paper.

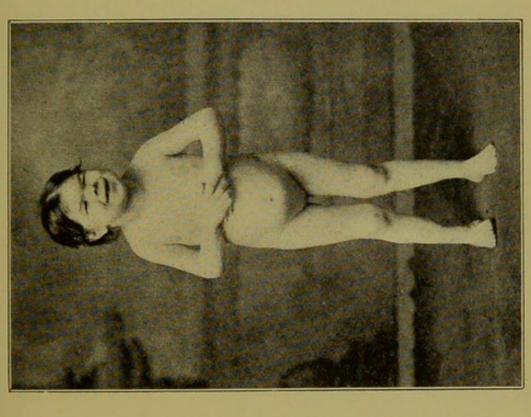


# JOURNAL OF MENTAL SCIENCE, APRIL, 1895.

PLATE II.

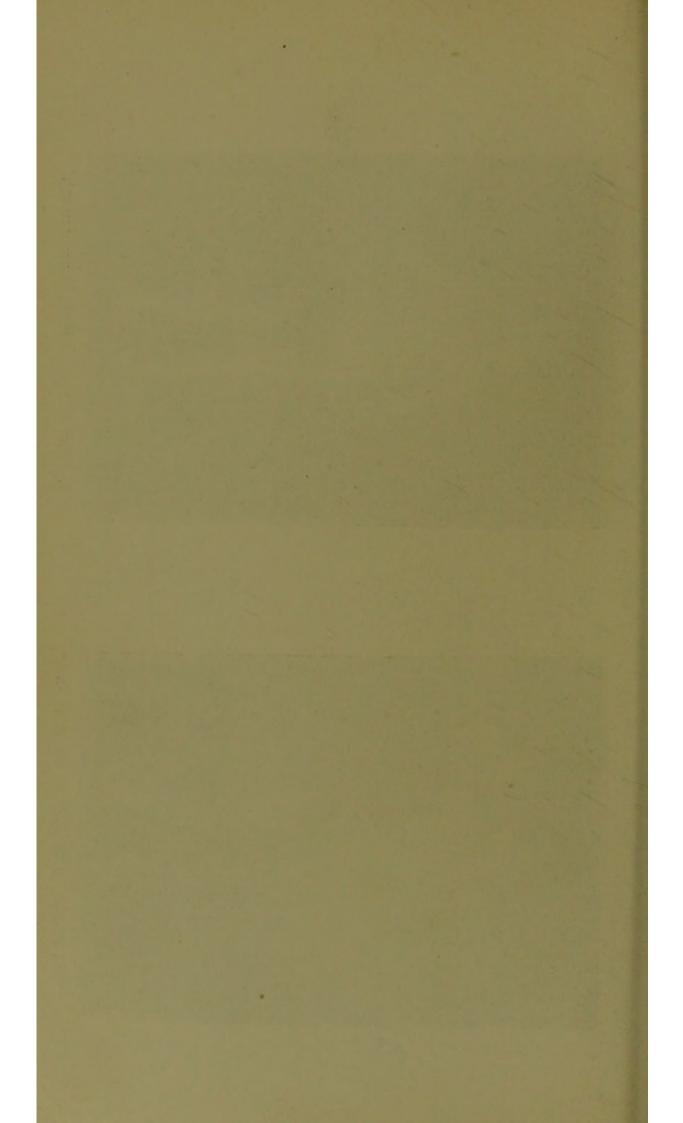


F. B. AT AGE OF 18 (BEFORE TREATMENT).



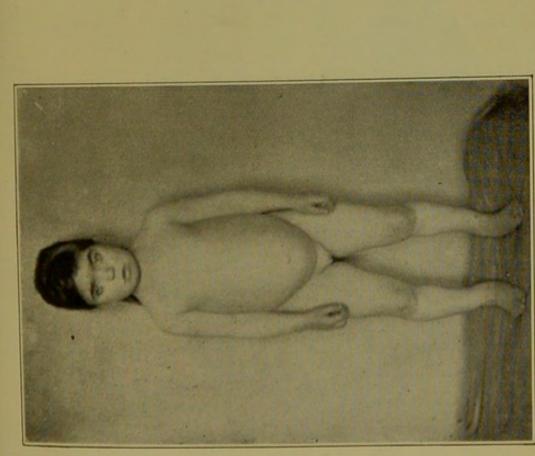
S. E. AT AGE OF 16 (BEFORE TREATMENT).

To illustrate Dr. Telford Smith's paper.

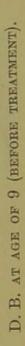


# JOURNAL OF MENTAL SCIENCE, APRIL, 1895.

PLATE III.



A. W. AT AGE OF 154 (BEFORE TREATMENT).

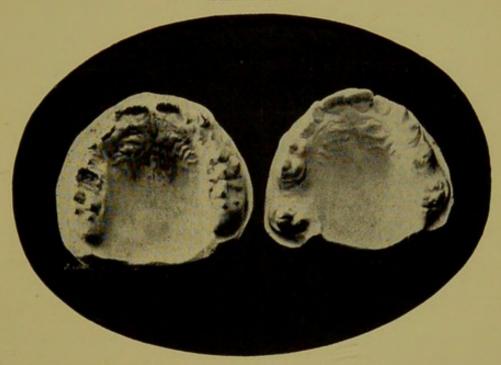


To illustrate Dr. Telford Smith's paper.



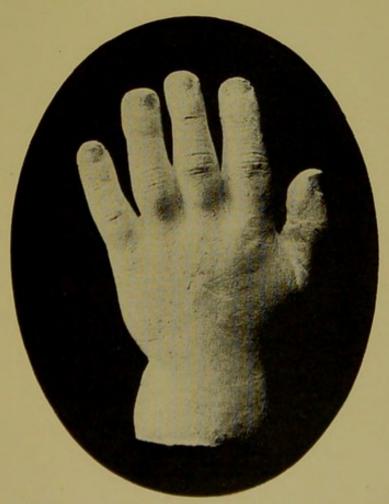
## JOURNAL OF MENTAL SCIENCE, APRIL, 1895.

PLATE IV.



PALATE OF S. E. F. B.

SHOWING CHARACTERISTIC SHAPE FOUND IN CRETINS—BROAD, FLAT.



HAND OF D. B. SQUARE AND SPADE-LIKE. (FROM CAST.)

To illustrate Dr. Telford Smith's paper.

## CASES OF SPORADIC CRETINISM TREATED BY THYROID EXTRACT.\*

BY TELFORD TELFORD-SMITH, M.A., M.D.,

Medical Superintendent, Royal Albert Asylum, Lancaster.

The four cases of sporadic cretinism the treatment of which by thyroid extract I shall briefly describe, are all

inmates of the Royal Albert Asylum, Lancaster.

They may, I think, be placed in Victor Horsley's third class of cases of cretinism, namely, that class in which the cretinism develops in early childhood, generally about or before the fifth year; that is to say, they are not cases of congenital cretinism, nor cases of ante-natal and subsequent slow development of cretinism. Three of the cases are girls and one a boy. They are all fairly typical, though not extreme cases, and the results of treatment by thyroid extract have been very similar in each case, though not equally marked in all.

Before treatment the general description of the sporadic

cretin applied to each of the four cases.

They were all dwarfed in size, and childish in appearance, with large heads and pale, waxy, and slightly yellowish complexions, and slight malar blush; the lips bluish and larger than normal, the lower protruding; the tongue more or less hypertrophied; the teeth defective and carious, and the eruption of the permanent set very much delayed; the palate wide and flat; the nose retroussé, very flattened between the eyes, and the nostrils wide and visible. The eyes widely separated, the palpebral fissure narrow, the eyes

<sup>\*</sup> Paper read by Dr. Shuttleworth, in the absence of the author, in the section of Psychology at the 62nd Annual Meeting of the British Medical Association, held at Bristol, July 31st, August 1st, 2nd, and 3rd, 1894.

almost disappearing when they smiled; the eyes normal and bright, the pupils reacting well to light and accommodation.

The wrists and ankles enlarged, and in the boy there is some beading of the ribs and thickening of the cranial sutures. The tibiæ are bowed and the feet flat. The hands short, thick and spade-like. They walk with a waddle, and can seldom run.

The deep reflexes are exaggerated in both extremities, and cutaneous sensibility almost absent in three of the cases.

The skin myxœdematous in appearance, rough and dry, perspiration seeming to be absent. The hair thin, short, dry, and growing very slowly.

The neck short and thick, and the thyroid gland either absent or hardly to be detected. Some fatty prominences

are to be felt in the supra-clavicular regions.

Their temperatures were all subnormal, averaging about

95-96°, and their extremities blue and cold.

Their bowels were generally constipated. Their abdomens were protuberant, and one of the girls and the boy had small umbilical herniæ.

They were extremely phlegmatic and slow in all their movements, even the expression of the face altering in a very gradual manner. They spoke very little, if at all, and in a thick and indistinct manner, using monosyllables. They understood fairly well what was said to them.

Their senses of sight, hearing, smell, and taste were, as

far as could be ascertained, about normal.

Their powers of observation, attention, and memory were

fairly good.

They were affectionate, but shy, and, as a rule, rather sulky and obstinate if annoyed.

Case F. B.—History of Patient.—She was born at Marcham,

Berks, 5th August, 1875 (now 19 years of age).

She is the second born child. She was born at full time and normally, but the labour was more difficult than with the first child, and was rather protracted. No instruments were used, and the child was not asphyxiated when born, nor was it convulsed soon after birth. She has had some convulsions during attacks of bronchitis, between her 5th and 10th years.

She did not begin to walk till about five years, and talking was

not attempted till about three years of age.

The mental deficiency did not begin to be noticed till about five

years, as she was a very fine baby and seemed quite right, except for the late walking and talking.

Family History of Patient.—The father is now aged 62, and is a native of Chester. He is a fine healthy man over six feet in height, and has been a gentleman's servant. He is normal in intelligence, is temperate; was not related by blood to his wife, nor is there any history of consanguineous marriage on either side.

There is no history of hereditary disease in his family, nor in that of his wife. No goitre; no nervous diseases.

The mother is a fine, healthy, intelligent woman, now aged 50. She is considerably over the average in height and physique, and is not neurotic. She says, however, that she was much worried and depressed during her pregnancy with the patient, through money losses.

M	EASUREN	ients (F. B.)			
Date.	Age.	Height. Inches.	Weight. lbs.		
1892, August	17	44	65		
1893, August	18	44	73		
September	Treat	ment commence	d.		
October	-	-	69		
1894, January	19	46	65		
March	_	463	70		
July	-	47	76		
		August, 1893.	July, 1894.		
		Inches.	Inches.		
Girth of Body round	Navel	. 28	261		
" " M	ammæ	. 283	28		

## HEAD MEASUREMENTS (F. B.) Inches. Circumference ... Above ears and over occipital tuberosity ... 20 Transverse ... (a) Tape measure ... ... ... 125 (b) Calliper measure from ear to ear over vertex ... Longitudinal... (a) Tape measure ... 12 (b) Calliper measure from nasal notch to occipital tuberosity ... Width of Forehead Width between inner Canthi of Eyes ... 18 outer ,, ,, ... 34 79.3 Cephalic Index ... Shape of Head-Brachycephalic.

Note, Nov., 1894.—This patient went to her home in Chester on August 6th, 1894, for a month's holiday. On September 2nd her father wrote to say that they would not send her back to the asylum as they thought she was too intelligent to be among our inmates.

CASE S. E.—History of Patient.—She was born at Huddersfield

11th January, 1877 (now 17 years of age).

She is the first-born child of the family, and was born at full time and normally. Labour was not protracted, nor were instruments used. She was not asphyxiated after birth, nor has she ever had any kind of fit.

Began to attempt to walk at about three years; she had a fall down some cellar steps when two years old, which kept her back.

She did not begin to talk till about seven years of age.

Her mother says she noticed some mental deficiency in the child from birth, and attributed it to being frightened during pregnancy by seeing a young man, an idiot, who lived next door, in epileptic fits. Family History of Patient.—Her father, now aged 52, is a coalminer, living at present at Barnsley.

He is a healthy and intelligent man. Has always been tem-

perate. No history of insanity or phthisis in his family.

Her mother is aged 52; is healthy and intelligent, and has always been temperate. No history of insanity or of phthisis in her family.

The father and mother were not related by blood, nor is there any history of consanguineous marriages in either of their

families.

There are two sisters younger than the patient, both healthy and intelligent.

HEAD MEASUREMENTS (S. E.)						
		1888. Inches.	1894. Inches.			
Circumference	Above ears and over occipital tuberosity	201	21			
Transverse	(a) Tape measure	111	123			
Longitudinal	(b) Calliper measure from ear to ear over vertex  (a) Tape measure  (b) Calliper measure from	5·5 12‡	5.65 121			
	nasal notch to occi- pital tuberosity	71	71			
Width of Forehead	4	41				
Width between inne		11/2				
,, ,, oute	r "		33			
Cephalic Index			77.9			
Shape of Head—Br	achycephalic					

Measurements (S. H.)						
Date.	Age.	Height. Inches.	Weight.			
1884, February	7	33½	_			
1889, July	11	41	48			
1892, April	15	44	54			
1893, March	Tre atment commen ced.					
April	16	441	531			
1894, July	17	46‡	58 <u>1</u>			
Ootober	-	49	66			
		March, 1893.	July, 1894.			
		Inches.	Inches.			
Girth of body round Navel		231	231			
" " Mammæ		231	241			

Case A. W.—History of Patient.—She was born 18th Novem-

ber, 1878, at Rochdale (now 15½ years).

She is the third-born child, and was born at full time and normally. Labour was not protracted nor difficult. No instruments were used. She was not asphyxiated when born, nor was she convulsed soon after birth. She has never had any kind of fit. Began to walk at about 12 months; was about six to seven years old before she commenced to talk. Her mental deficiency began to be noticed at about one year. Her mother attributed it to fretting during her pregnancy with patient, owing to the eldest daughter being "sent away."

Family History of Patient.—Her father, now aged 56, is a coalminer, living in Rochdale; is a strong and apparently healthy man, but is intemperate. His niece has epileptic fits, but is at

work.

The patient's mother is a strong, temperate woman, aged 44, who had an aunt insane for 20 years. There is no his-

tory of consanguineous marriages in the family. No history of

phthisis.

The eldest sister of patient died in Whittingham Asylum in 1893, aged about 20. She is said to have had epileptic fits after about two years of age, due to having been bitten by a dog.

There are four younger children in the family, three girls and

one boy; normal in physical and mental condition.

	F	IEAD	MEASUI	REMENTS	(A. V	V.)			
									Inches.
Circumference		Abo	ve ears	and ove	r occipi	ital tu	berosit	y	21
Transverse		(a)	Tape m	easure			F		121/2
		(b)	Calliper verte	measui x		ear	to ear	over	41
Longitudinal		(a)	Tape m	easure					131
		(b)	Calliper	measu					71
Width of Forehe	ad								5
Width between i	nner (	Cantl	ni of Ey	es				***	14
" "	outer	"	,,						33
Cephalic Index									73.4
Shape of Head-	-Dolio	choce	phalic.						

Case D. B.—History of Patient.—The patient was born on June 24th, 1884. He is the fourth-born child. His eldest brother, aged 14, is also a cretin. A healthy brother and sister were born between the patient and his eldest brother. There are five brothers and sisters healthy and normal, physically and mentally. D. B. was born at full time and normally; labour was protracted. No instruments were used, and he was not asphyxiated when born. He was not convulsed soon after birth, nor has he ever had any kind of fit. He is said to have been bright as an infant, and cut his first teeth early. His mental deficiency began to be noticed at about two years of age. He did not begin to walk till about  $2\frac{1}{2}$  years. He has had measles, whooping cough, and diphtheria.

Family History of Patient.—His father, aged 45, is a fish salesman in Manchester. He is temperate and is an intelligent man. He is stout and measures about 5ft. 6in. in height. Is subject to asthma, as was his (the father's) mother. There is no history of intemperance in his family. The patient's mother is aged 35. She is temperate and an intelligent but very neurotic woman, and inclined to be delicate. Her father was intemperate, and died at the age of 44, of phthisis. There is no further history of intemperance in her family. The father and mother of the patient were not related, and there is no history of consanguineous marriages in their families. There is no history of insanity on either side.

Note, Nov., 1894.—The boy D. B. went to his home in Manchester on Aug. 6th, 1894, for a month's holiday, which was extended on account of infection in the house. On Oct. 23rd, 1894, his mother wrote to say they had now made up their minds to keep him at home, "as he has come on better than we expected when he first went to the asylum, and he is getting on splendid at home, now he goes to school regular weekdays and Sundays, and also to church, and he has brightened up wonderful, and he knows several tunes o songs and hymns that he hears at school and church, and h knows the chorus of 'Daisy Bell.' He is two inches taller and i half a pound heavier than when he came home."

Through the kindness of Dr. R. Turner, of York, I am permitte to make use of his notes on the blood of two of the cases (females of cretinism mentioned in this paper, the examination having been made by Dr. Cattley, of York, on some blood he took from

the patients in the Royal Albert Asylum.

Dr. Turner says (in his paper on "Cretinism and Myxœdema," read before the York Medical Society, October, 1893). "It is an interesting fact to note that the pathological changes in the blood in cretinism are identical with the pathological changes in myxœdema. Apart from the diminution in the red cell element and an increase in the leucocytes the two principal and characteristic features are the following:—

I. The Presence of Marrow Cells in the Blood.—These cells are large, and stain a light Cambridge blue with hæmatoxylin, but

are not in any way affected with eosin.

The nucleus of the cell is only of a slightly deeper colour than the protoplasm, and it is quite unlike the violet tint of the multinucleated cells. The nucleus is generally identical on one side.

II. The Large Number of Eosinophile Cells.—In these the nucleus has the same blue tint that we find in the nucleus of the marrow cells, but the protoplasm contains numbers of bright spherical refracting granules which stain with eosin.

Each of these patients has been under treatment for different periods, and are all at present taking one 5gr. tabloid (Burroughs and Wellcome) daily at dinner time.

The improvement is well marked in each. The chief effects of the treatment may be summed up as follows:—

Almost after the first dose the temperature began to rise, gradually approaching the normal as treatment was continued

At about two to three weeks the skin began to desquamate, chiefly on the hands, feet, and face, and to assume a more normal tint and feel; it lost its dry, rough, waxy appearance,

and perspiration became perceptible.

The myxœdematous condition of the subcutaneous tissues began to subside, and the outline of the features became more defined and sharp. The abdomen became less protuberant, the hands and feet less thick and spade-like, and there was a general loss of weight, which, if the dose of thyroid was excessive, verged on a state of emaciation, the ribs becoming visible and the general nutrition evidently impaired. If, however, the dose was kept at a minimum the general muscular nutrition seemed to improve, and the previous loss of weight to be again made up by healthy tissue. A comparatively rapid increase in height commenced, and the previously delayed second dentition began to appear. The cutaneous sensibility became more near the normal, and the marked constipation gave place to a more healthy regularity. In the two cases where an umbilical hernia was present it became almost imperceptible.

After about two or three weeks' treatment a gradual change became noticeable in their temperament and manners; the phlegmatic state gave place to a spontaneous activity, their expression became more lively and intelligent. They became playful and even mischievous, and were constantly in motion, and the sulkiness of temper and

disinclination to be amused passed off.

In the course of the treatment of these cases I had occasion to suspend the administration of the thyroid for several months. During this time I found that the patients slowly and gradually reverted to their former condition of mind and body, but this lapse was much more gradual than their former improvement had been. I should say that the deterioration occupied more than twice the length of time which was needed for the improvement.

The two conditions—the temperature at night, together with the state of general nutrition—afford a useful guide as to when a sufficient dose of the thyroid is being administered. If the temperature can be kept at about 97.5 to 98

Fah., and at the same time no emaciation is set up, the physiologically useful dose has been gauged. But if flesh is steadily lost and any degree of emaciation is set up, the dose should be diminished, even if the temperature should still remain considerably below normal.

In commencing treatment it is well to begin with a small dose—say 3grs. a day—and to increase it gradually to five

or more grains, according to the effect produced.

If a large initial dose is given symptoms of great depression may show themselves—vomiting, headache, cold sweats, fall of temperature, followed by rise to 103 Fah. or higher,

and signs of heart failure.

Judging from the different degrees of improvement in these cases, and in others that have been published, I think the conclusion that "in cases of equal degree the younger we can commence treatment the greater will be the improvement" is borne out.

And one is led to the speculation whether if we could detect a case of sporadic cretinism or cretinoid imbecility in its very early stage, when the child was in its infancy and before its mental and physical constitution had become impressed with the, I fear, not entirely eradicable stamp of the disease: whether in this early stage, if treatment were commenced and the physiological defect supplied by thyroid administration, the child might not grow up in an almost normal condition of mind and body. This early detection and treatment of the disease rests largely with the family doctor, as the parents are not so likely to notice or acknowledge the gradually increasing mental deficiency of their child, and the cretinoid condition is probably well-established before the case is brought for special care.

This form of imbecility certainly appears to be the one which offers the most hope of improvement from early and continuous medical treatment, apart from special training.

I am glad to be able to report that the improvement in the four cretinoid patients treated with Thyroid Extract continues.

Two of these patients, a boy and a girl, have been withdrawn from the Asylum during the year, their parents considering that they were now "too good" to remain among I have lately written to the parents, our other patients. asking for information regarding their present condition, and have had a reply from the mother of the little boy, in which she says, "D. has grown 4 inches in height since he came home last August (he is now 421 inches), he is very active on his feet, and he has improved in his talk, he attends school, and the Head Schoolmistress tells us he has made very fair progress, he is able to do little lessons by dictation, he has learnt tunes of hymns and songs, and learns a good many of the words in the hymns. Dr. Shuttleworth came to see him a fortnight ago, and said he had grown a bonny boy,"

If we recall the boy's condition when he came to the Asylum—a stunted dwarf of 33 inches in height, with little prospect of further growth; almost speechless; apathetic, sitting motionless for an indefinite time, and taking no interest in his surroundings—and contrast it with his present state, the result is certainly remarkable and most encouraging.

\*\*RAA Relate 1895\*\*

## THYNCE EATRACT FOR BACKWARD CHILDREN.

The use of thyroid extract in the treatment of sporadic cretinism is of course well known, but Dr. Clement Dukes, physician to Rugby School, suggests its employment as an aid in the development of children who are merely backward. He relates a case in which, although the child looked younger than her age, she was well-formed and intelligent, ate, drank, and slept well, was very well nourished, took her part both at home and at school the same as other children, and was like the other members of her family, except that she was short in stature and small made, and had a pallor which neither arsenic or iron had ever remedied. giving thyroid tablets (one of five grains once a day, and after a month twice a day) her pallor disappeared, she became much more brisk and lively, and started growing; in fact, she became a different child altogether. This is certainly a case worthy of note, an experiment Hosp: 12.3.98 worthy of repetition.

in the case of their children.

As far as can be learned there are no cases in which any material benefit has resulted after the operation, although the total of cases operated on in France, in America, and in this country probably numbers two hundred.

This year I had the honour of being associated with Professor Cunningham, of Trinity College, Dublin, in publishing a memoir on "The Brain of the Microcephalic Idioth in the Scientific Transactions of the Royal Dublin Society and, after an exhaustive anatomical examination of such brains, this strong opinion is expressed regarding the operation of Craniectomy, "We have no hesitation in saying that it would be quite as rational a proceeding to operate on the head of an ape in the hope of producing an access of brain growth as upon the head of a typical case of microcephaly.

In conclusion I would beg leave to acknowledge the zealous and efficient aid of my colleague Dr. A. R. Douglas in all that tends to promote the health and welfare of the patients.

I have the honour to be, Gentlemen, Your obedient Servant,

T. TELFORD-SMITH,

Medical Superintendent.

per cent. of the whole number, with a is diagnostic of in the neutrophiles. Whether this is diagnostic of

trichinosis remains to be seen. Though the discovery of bacteria in the blood is too uncertain to be of much value for the diagnosis of the disease, a positive result shows that more than a local infection has occurred, and that a general septicæmia has taken place. Kohn's results in pneumonia are the most interesting of the numerous investigations he made. Out of 32 cases of pneumonia he found the diplococcus in the blood in nine, seven of which were fatal, and the other two presented secondary abscesses; while, on the other hand, no death from pure pneumonia took place in the patients whose blood was free from the coccus. He therefore looks on the appearance of cocci in the blood as of the worst augury, and as generally indicating a septicæmic condition or multiple local centres of the poison. Since blood serum has been so often tested as a means of diagnosis, those cases where it is cloudy or turbid have acquired! additional interest. Castaigne finds that the turbidity