

SYPHILIS IN MENTAL DEFICIENCY*

BY

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In the course of examining children with congenital syphilis, Paddle (1940) noted that 7 per cent. were mental defectives; the incidence of congenital syphilis amongst mental defective children has been variously assessed at 16.5 per cent. by Gordon (1913) 25 per cent. by Stewart (1926), and 4.3 per cent. by Benda (1942); and Dean (1912) observed that 16.1 per cent. of idiots suffered from inherited syphilis.

Equally conflicting have been the results of attempts to determine the part played by syphilis in producing mental deficiency. Jeans and Butler (1914) estimated the incidence of mental deficiency to be five times more common in congenital syphilitics than in the general community. According to Stokes and others (1944), Benda and Tadgell (1942) suggested that 40 to 50 per cent. of the feeble-minded with congenital syphilis owe their mental deficiency to this disease, an opinion shared by Fraser and Watson (1913). Lloyd (1943), forming his opinion upon a very low serological incidence of syphilis (0.78 per cent.) in juvenile defectives whose average age was 5½ years, did not agree.

At the request of the medical superintendents in charge of two large colonies for mental defectives in the North-East of England, a total of 1,331 patients were examined clinically and serologically with a view to establishing the incidence of syphilis.

Syphilis was found in 2.82 per cent. of the feeble-

minded and in 1.86 per cent. of imbeciles, but not in the idiots. One female belonged to the 6-10 age group, one male and two females were between the ages of 11 and 16, and the remaining thirteen males and fifteen females were at least 17 years of age (Table I).

Serological Diagnosis

Serum Wassermann tests were performed at one of two local laboratories, all initial positive or doubtful results at each being repeated at the other. Both laboratories employed the quantitative Wassermann reaction, consisting of serial dilution of the patient's serum from 1:2 to 1:32 with a fixed complement dosage throughout. Repeated strongly positive Wassermann results were necessary before a diagnosis of syphilis was made in the absence of confirmatory clinical evidence or past history.

Of 1,331 blood specimens subjected to Wassermann examination, 31 were initially positive or doubtful. On repetition, 24 (1.8 per cent.), again proved to be strongly positive, five were found to be negative, and two who maintained a low titre were eventually proved to be free from syphilis (Table II, opposite).

A comprehensive survey embodying the statistics recorded in this and 28 other publications showed that of a total of 21,002 mental defectives in Gt. Britain and abroad, 1,365 (6.5 per cent.) had sero-

TABLE I
AGE CLASSIFICATION IN RELATION TO TYPES OF MENTAL DEFICIENCY AND INCIDENCE OF SYPHILIS
(IN PARENTHESES) IN 1,331 PATIENTS

Age Group	Sex	Feeble-Minded	Imbecile	Idiot	Mentally Defective		Syphilitic																																												
					Total	Percentage	Total	Percentage																																											
-5	M	2	3	11	16 } 2 }	1.4	—	—																																											
	F	1	1	—					6-10	M	15	14	12	41 } 29 }	5.2	1	1.4	F	13	14 (1)	2	11-16	M	51 (1)	22	12	85 } 51 }	10.2	3	2.2	F	25 (1)	23 (1)	3	17+	M	462 (11)	80 (2)	44	586 } 521 }	83.2	28	2.5	F	388 (14)	112 (1)	21	Totals		957 (27)	269 (5)
6-10	M	15	14	12	41 } 29 }	5.2	1	1.4																																											
	F	13	14 (1)	2					11-16	M	51 (1)	22	12	85 } 51 }	10.2	3	2.2	F	25 (1)	23 (1)	3	17+	M	462 (11)	80 (2)	44	586 } 521 }	83.2	28	2.5	F	388 (14)	112 (1)	21	Totals		957 (27)	269 (5)	105	1,331	100	32	2.4								
11-16	M	51 (1)	22	12	85 } 51 }	10.2	3	2.2																																											
	F	25 (1)	23 (1)	3					17+	M	462 (11)	80 (2)	44	586 } 521 }	83.2	28	2.5	F	388 (14)	112 (1)	21	Totals		957 (27)	269 (5)	105	1,331	100	32	2.4																					
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TABLE II
 GROUNDS FOR DIAGNOSING SYPHILIS

No. of Patients	Serum Wassermann Reaction	Clinical Manifestations	Cerebro-spinal Fluid Results	Past History (family/patient)
10	+	+	—	—
7	—	—	—	—
6	++	—	—	—
3	++	+	+	+
2	++	—	—	—
1	++	+	—	+
1	—	—	—	+
1	—	—	—	+
1	+	—	—	+

logical evidence of syphilis. A wide divergence in results was noted. For example, Lloyd (1943) observed 0.78 per cent. positive results compared with 55.6 per cent. found by Key and Pijper (1922). Our figure of 1.8 per cent., which is more in keeping with Lloyd's findings and is well below the mean, may have been due to modern serological complement-fixation tests presenting increased specificity as well as sensitivity. Again, the flocculation tests employed by some authors, are probably more sensitive than complement-fixation tests but the selectivity is suspect, e.g., the Kahn presumptive and Kline exclusion tests yielding an established margin or error up to about 10 per cent. Nor must we overlook the decline in the incidence of syphilis in certain population groups when attempting to explain the divergence between our figures and those obtained in past years by other authors.

The age of the patients under review is also an important factor and one to which we are inclined to attach more importance than to any other. Especially is this the case in congenital syphilis where serological evidence of the disease varies inversely with the age of the patient. Churchill (1912) found a 38 per cent. incidence in 102 infants and children, and Fraser and Watson (1913) found no less than 46.3 per cent. of 205 mental defectives under the age of 17 years to have a positive serum Wassermann reaction. It will be seen from Table I that, owing to the great preponderance of "over 17s" (83 per cent.) in our survey, the prospect of detecting congenital syphilis by serological tests was considerably lessened. In an attempt to link up the prevalence of the disease not only with the age of the patient but also with the sex, Paddle (1934) noted that the incidence graph was at its highest in childhood in both sexes, and at its lowest in women between 31 and 40 and in men between 41 and 50, thereafter showing a rise in both sexes. It is suggested that the widely divergent results arising from serological surveys amongst mental defectives might therefore be due to a combination of factors.

Clinical Diagnosis

Eventually 32 (2.4 per cent.) of our patients were found to be syphilitic, 21 revealing clinical evidence of the disease. Ten presented classical manifestations: Hutchinsonian teeth—four, Moon's molar—one, and Argyll-Robertson pupils—five, in accordance with the definition of Prof. F. J. Nattrass (see below). Other dental abnormalities of secondary importance, epilepsy, and the various plegias were also noted amongst those infected.

The limited value of the serological approach as a diagnostic measure is illustrated by the observation of Woodall (1929) that fully 50 per cent. of untreated congenital syphilitics become serologically negative with the passage of time. Again, since little or no reliance can be placed upon the case history given by the mental defective, a negative serology may considerably magnify the importance of clinical findings in such a patient. Even so, clinical criteria, which at first glance might be regarded as diagnostic, are not always recognized as pathognomonic of congenital syphilis. Two examples may be given:

(a) Some authorities (e.g. Purves-Stewart, 1924) maintain that Argyll-Robertson pupils occur in alcoholism, syringo-myelia, and other conditions independent of neurosyphilis. All would appear to agree that such pupils do not react to light and do react normally to accommodation, but if we extend the definition to include miosis only, as suggested by Nattrass (1951), then it is probably reasonable to assume that all such pupils are of syphilitic origin.

(b) In the medical literature, reference is periodically made to Hutchinson incisors caused by factors or diseases other than inherited syphilis. Such a view (for example that held by Ruiz Santamaria, 1949) is regarded as untenable by Prof. R. V. Bradlaw and Prof. J. Boyes of the Newcastle-upon-Tyne Dental School.

Other clinical evidence, less dogmatic diagnostically, occurs with considerable frequency. Thus, interstitial keratitis in the absence of corroborative evidence cannot always be ascribed to inherited syphilis and may be due to tuberculosis, focal infection, malaria, or myxoedema, to mention only a few aetiological factors. Amongst the 1,331 mental defectives under review, syphilis was found in one who had a past history of interstitial keratitis but it could not be detected in twenty others presenting corneal opacities.

Other ocular manifestations such as irregular or unequal pupils, were observed in 126 (9.47 per cent.) patients in our series, of whom seven were proved to have syphilis; twenty (1.5 per cent.) exhibited pupils which failed to react to light but were not regarded as Argyll-Robertson pupils, and syphilis

was absent in 81 (6.1 per cent.) patients showing strabismus, and in six with ptosis.

Eighty-nine (6.68 per cent.) patients had a history of epilepsy and 42 of them presented no other clinical abnormality. Six (7 per cent.) epileptics were syphilitic and to these may be added four others who were serologically negative but presented clinical features of admittedly lesser diagnostic importance (for example, corneal opacities). There was no evidence to show that epilepsy was due to syphilis in any of the six patients, an observation in accordance with the findings of Gordon (1913) and Paddle (1934).

Paralysis involving one or more extremities was noted in 69 (5.2 per cent.) patients, of whom three (4.4 per cent.) suffered from syphilis. Gordon (1913) found amongst his plegias that 31.4 per cent. had positive blood Wassermann reactions as compared with 11.2 per cent. of his non-plegic patients.

Of nine patients who were deaf and dumb, none had syphilis. Thomsen and others (1911) noted a positive serology in 0.87 per cent. of 344 such patients.

Optic atrophy accounted for the only blind patient found to have syphilis. Again we quote Thomsen and others (1911) who found no evidence of syphilis in 146 blind patients examined.

Amongst 32 syphilitic mental defectives, 27 were feeble-minded and the remainder were classified as imbeciles. This classification is at variance with the observations of Stewart (1926) and Woodall (1929), who found the highest incidence of syphilis amongst imbeciles and not amongst the feeble-minded. Perhaps the diversity in results can be attributed to the fact that in our series syphilis was incidental to rather than the cause of mental deficiency.

Cerebrospinal Fluid Investigation

Thirty-one infected patients were subjected to cerebrospinal fluid investigation, diffuse spinal osteo-arthritis preventing lumbar puncture in one woman. Examination included cell count, globulin estimation, quantitative Wassermann reaction and gold sol test. A diagnosis of neurosyphilis was made in six (19.5 per cent.) patients, of whom three presented no clinical evidence of the disease. The remaining three, one of whom had classical Hutchinsonian teeth, were epileptics.

Congenital Syphilis

Striking clinical manifestations, such as Hutchinsonian teeth (4), Moon's molars (1), Argyll-Robert-

son pupils (2), were noted in six out of a total of ten patients who had congenital syphilis. Such clinical manifestations occurred alone or in conjunction with one or more of the other diagnostic criteria: positive serological evidence (5), previous medical history (1), youth of the patient (4). Four patients had active spinal fluids. This is in accordance with the observations of Paddle (1934 and 1940), who remarked upon the incidence of abnormal cerebrospinal fluids varying between 21 and 33 per cent. Eskuchen (1919) and Greenfield and Carmichael (1925) noted the frequency of atypical cerebrospinal fluid findings in congenital syphilis, except in the classical juvenile parietic and tabetic pictures.

Family History and Previous History of the Patient

A family follow-up was feasible in only fifteen patients since the majority of the remainder resided in districts outside the jurisdiction of this clinic. Twenty parents had died, two were positive when examined, three showed no evidence of the disease, and advanced senility precluded the possibility of examination in the remaining five.

Two sisters were found to have congenital syphilis, seven proved negative on examination, two refused to cooperate, and a further two could not be located.

Of nineteen brothers, one was found to have serological syphilis, sixteen were negative, and two failed to cooperate.

The patients themselves (of whom seven were known to be sexually promiscuous), or their parents on their behalf, admitted to nine illegitimate children, of whom five died at birth or shortly afterwards, three could not be traced, and the only one examined was found to be free from syphilis.

The past case history of the patient was of value in three males only: one, aged 23, yielded a history of interstitial keratitis at the age of 14 years; another, aged 49, was found to have received treatment previously in this department for tertiary lesions; the third, aged 14, had previously been treated for congenital syphilitic infection.

Summary

Out of 1,331 mental defectives examined for syphilis, 32 (2.4 per cent.) were judged (principally on clinical and serological grounds) to have been infected. The past histories of the patients and their families were also considered.

Serological evidence formed the most reliable grounds upon which diagnosis could be made.

Classical congenital stigmata were present in only six patients; altogether, ten were definitely

proved to have the congenital variety of the disease, but in some of the remainder, it must be acknowledged that syphilis might have been inherited.

There was no reason to suppose that syphilis accounted for mental deficiency to any real extent.

We would take this opportunity of recording our grateful thanks to Doctors G. McCoull and C. G. Millman, Medical Superintendents in charge of Prudhoe and Northgate Mental Deficiency Hospitals respectively, but for whose cooperation and invaluable assistance this investigation would not have been possible.

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