THE INCIDENCE OF PORPHYRIA CUTANEA TARDA IN THE BANTU

AN ANALYSIS BY SEX AND AGE

K. J. KEELEY, M.B., B.CH. (RAND), M.R.C.P. (EDIN.)

Department of Medicine, Baragwanath Hospital and the University of the Witwatersrand, Johannesburg

The common occurrence of porphyria cutanea tarda (PCT) in the Bantu population has been stressed by Barnes.[1] By 1955 he had demonstrated porphyrin in the urine of nearly 300 non-Europeans, mostly Bantu. He also recorded the age and sex distribution in a series of over 100 cases; these patients were, however, derived from a selected hospital population. Findlay[2] reported that PCT accounted for 1% of patients at a Bantu dermatology clinic. Information has been lacking regarding the incidence in a less selected population group. The object of the present study was to establish the incidence of PCT with particular reference to age and sex.

MATERIAL

The main survey was conducted in the outpatient department of Baragwanath Hospital in 1960. Urine samples, to be tested for porphyrin, were collected from all patients awaiting attention. These patients included all those with non-acute surgical and medical complaints; those too ill to sit on the benches were automatically excluded. Many had trivial complaints which, in a White population, would not have led to attendance at a hospital. This group was considered a reasonably good one for study despite the limitations inherent in surveying a hospital population, even an outpatient one.

METHODS

The first 1,000 consecutive specimens of urine were collected without any selection, except that obviously blood-stained specimens were rejected (Table I). The imbalance between the sexes and between the older and younger age groups was such, however, that further collections

TABLE I. INCIDENCE OF PORPHYRIA IN AN UNSELECTED AFRICAN POPULATION

Age group (years)	*Male*	*Female*	*Total*
10 — 19	0 : 70	0 : 126	0 : 196
20 — 29	0 : 83	0 : 246	0 : 329
30 — 39	0 : 97	2 : 136	2 : 233
40 — 49	1 : 70	1 : 69	2 : 139
50 — 59	2 : 38	1 : 26	3 : 64
60 + ...	1 : 22	0 : 18	1 : 40
Total	4 : 380	4 : 621	8 : 1,001

of urine were made in stages from: (*a*) additional males of all ages, (*b*) both sexes over 40 years of age, and (*c*) both sexes over 50 years of age.

All specimens were examined promptly, and I personally performed the spectroscopy. Definitely and suspiciously positive specimens were examined microscopically for red cells, and by repeat spectroscopy after the addition of concentrated hydrochloric acid.

By the spectroscopic method as little as 50 - 100 μg. per 100 ml. of porphyrin may be detected, depending on the experience of the observer;[3] since the excretion of uroporphyrin does not normally exceed 30 μg. per 24 hours, the method errs on the side of undersensitivity; indeed, in the majority of positive cases the colour presented by the specimen to the naked eye arouses suspicion.

All but 3 subjects giving a positive result were examined clinically for signs of PCT. Attempts by a field worker to trace these 3 persons were unavailing.

RESULTS

A positive result was obtained in 25 specimens. Two patients were excluded from the series — Bantu women of 34 years and 52 years, who complained of facial hirsuties and crusts on the hands, respectively, and were thus regarded as self-selected. There remained 23 persons who had attended because of unrelated complaints (Table II); of these 20 were available for clinical examination, and all but 3 of them were recognizably porphyric (Table III). The 3 showing no evidence of PCT were regarded as latent cases.

TABLE II. INCIDENCE OF PORPHYRIA IN AN ADJUSTED AFRICAN POPULATION

Age group (years)	*Male*	*Female*	*Total*	*%*
10 — 19 ..	0 : 114	0 : 126	0 : 240	—
20 — 29 ..	0 : 155	0 : 246	0 : 401	—
30 — 39 ..	1 : 186	2 : 136	3 : 322	0·93
40 — 49 ..	5 : 226	2 : 125	7 : 351	1·99
50 — 59 ..	6 : 181	5 : 109	11 : 290	3·79
60 + ..	2 : 93	0 : 66	2 : 159	1·25
Total ..	14 : 955 (1·46%)	9 : 808 (1·11%)	23 : 1,763 (1·30%)	

Confirmation of the diagnosis was obtained in 2 cases, 1 latent and 1 overt, by the quantitative estimation of porphyrins. These results, together with those of other subjects, are given in Table IV.

In 10 of the 23 patients the urine was tested for sugar; in all 10 this test was negative. In a further patient, Julia K., glycosuria was detected in 1961 when she was admitted for pleurisy.

PENSIONER SURVEY

Because few elderly people attend the hospital, a complementary survey was carried out on a group of old-age pensioners who were being examined as a routine in the course of an unrelated clinical research project. All were over 60 years of age; there were 4 positive results in 230 women examined, 2 of whom had no clinical evidence of PCT. Only 1 male of the 83 examined was positive; he was a 66-year-old diabetic. The results of a quantitative examination are seen in Table IV. Thus, 1·59% of this

TABLE III. CLINICAL FINDINGS IN 23 SUBJECTS

Subject	*Age*	*Complaint*	*Evidence of porphyria*			
			Bullae	*Scars*	*Hyperpigmentation*	*Hirsuties*
James K. ..	59	Intermittent claudication	o	+++ Face and hands	Face	o
Clarina M. ..	37	Upper resp. infection	o	++ Hands	Face and hands, slight	o
Doris M. ..	40	Upper resp. infection	o	+ Hands	Dark facial	o
Lizzie D. ..	37	Headache	o	+ Hands	Peri-orbital	+
Robert T. ..	48	Fractured metatarsal	1958	o	Generalized facial	Malar
Union D. ..	60	Emphysema	o	+ Hands	Facial	o
Sarah N.* ..	52	Impetigo of ears	o	o	o	o
David M. ..	54	Hypertensive heart disease	o	o	Generalized facial	o
Joseph M.**	43	Asthma	—	—	—	—
Isaac Q. ..	30	Disseminated tuberculosis	o	o	Face	+
Bellina R. ..	50	Upper resp. infection	o	o	Peri-orbital	Slight facial
James M. ..	56	Carcinoma of oesophagus	1941	++ Hands	Generalized facial	o
Botha M. ..	45	Carcinoma of mouth	o	o	Generalized	o
Michael M.*	49	Trauma to face	o	o	o	o
George N. ..	55	Spondylitis	o	o	Face	Malar
Anna T.** ..	59	Upper resp. infection	—	—	—	—
Cornelius M.	49	Upper resp. infection	o	Minute on hands	Forehead and nose	o
Julia K. ..	59	Rash	o	+ Hands. + Feet	o	Frontal
William G. ..	60	Asthma	o	o	Peri-orbital	o
William N.*	50	Lung abscess	o	o	o	o
Anna M. ..	44	Upper resp. infection	History of bullae	o	Patchy facial	o
Elias M. ..	53	Fractured ulna	o Recent sores	+ Hands	o	o
Martha L.**	57	Upper resp. infection. Hypertensive	—	—	—	—

* Latent cases.
** Not examined.

TABLE IV. EXCRETION OF PORPHYRINS IN THE URINE IN μG. PER 24 HOURS

Series	*Name*	*Uroporphyrin*	*Coproporphyrin*
Outpatient ..	William N.*	1,300	195
	Elias M.	440	71
Pensioner ..	Jacob	2,980	414
Diabetic clinic ..	Isaac P.*	290	165
	Robert K.	4,350	604

* Latent cases.

age group were positive compared with 1·25% of a smaller number in the same age group in the outpatient population.

DIABETIC-CLINIC SURVEY

The final survey was made in the diabetic clinic to compare the incidence in this special group with that established for the general population. The results are given in Table V.

Because of the small number examined the contribution of age is difficult to evaluate; it seems likely, however, that the incidence in a diabetic population is higher than in the general population, and this is particularly noteworthy in the fourth decade.

Two of the positive subjects had latent PCT, and a quantitative estimation was done in one of these (Table IV). The overall incidence of 4·0% is higher than that in the general outpatient population (1·3%) and far higher than recorded in diabetic populations in Europe. In 2 recorded series of diabetics in Europe,[4,5] although both

TABLE V. COMPARISON OF INCIDENCE OF PCT IN DIABETIC AFRICANS AND IN THE GENERAL AFRICAN POPULATION

Age group (years)	*Ratio expected**	*Ratio found*	*No. found*	*%*
All ages	1 : 76	1 : 25	6 : 150	4·0
30 — 39	1 : 107	1 : 7	3 : 23	
40 — 49	1 : 50	1 : 36	1 : 36	
50 — 59	1 : 26	1 : 24	2 : 48	
60 + ...	1 : 79	—	0 : 28	

* Calculated from Table II.

sexes were investigated, porphyria was found only in the males, 7 in 1,602 cases (0·43%), and 3 in 450 cases (0·66%), respectively.

CONCLUSIONS

1. In the general population there appeared to be a rising incidence with increasing age, reaching a peak in the sixth decade; the majority of cases occurred between the ages of 40 and 59 years. The incidence in pensioners was much the same as in the 60+ division of the general population. By contrast, the peak incidence in the diabetic group was in the fourth decade.

2. The incidence in males and females did not differ significantly.

3. Glycosuria was not detected in porphyrics from the general population, but was present in 1 porphyric pensioner. The incidence of porphyria in the diabetic clinic was remarkably high.

4. Latent cases were found surprisingly often; the ratio of latent to overt cases was as follows: general population — 3:20; pensioners — 2:3; and diabetics — 2:4.

DISCUSSION

Reports in the literature on the sex ratio in PCT have been confusing: these vary from a male predominance

of 3·2:1 and 4·7:1 in American white populations,[6,7] to 2·6:1 in favour of females in Barnes' Bantu series.[1] My own experience of patients admitted to a medical unit confirmed Barnes' impression. However, Lamont *et al.*,[8] in 100 cases, found a slight excess of males, 1·4:1, in a Durban Bantu population.

The present survey suggests that there is no significant difference in incidence in the sexes in an urban outpatient population. Two factors probably contribute to the number of female porphyrics admitted; firstly, PCT is more readily diagnosed in women because hirsuties is more obvious in this sex, and hyperpigmentation is more disturbing to them; secondly, they are freer to attend the hospital than the working males. These suggestions are supported by the number of female attendances shown in Table I; it is likewise noteworthy that the 2 subjects excluded from the survey were both women complaining of the skin lesions of PCT.

In both previously reported Bantu series mentioned above,[1,8] the majority of subjects were aged between 30 and 50 years; it was surprising therefore to find that the majority in the general population sample in this series were between 40 and 59 years and that the peak incidence occurred in the sixth decade. This is probably explained by the fact that care was taken to include sufficient numbers of elderly people in the present study. A more rapidly progressive process may be present in those patients in whom porphyria and diabetes were associated, the peak incidence being in the fourth decade. These 2 metabolic anomalies may have a common cause, such as oral-iron overload; the target organs, the liver and the pancreas, may vary in sensitivity so that glycosuria may precede or follow porphyria. Alternatively, 2 or more noxious agents, such as iron and ethanol, may be associated, as in the illicit brews drunk by the Bantu.

The extraordinarily high incidence of porphyria in Bantu diabetics is partly explained by the fact that the incidence in the general Bantu population exceeds that in European diabetics, and possibly by the fact that 7% of Bantu diabetics have siderosis of a haemochromatotic distribution.[9]

Attention should be paid to the occurrence of latent cases, that is, those in which excessive amounts of uroporphyrin and coproporphyrin are excreted in the urine in the absence of clinical signs. Barnes[1] referred briefly to 2 possible latent subjects, both of whom had sibs with overt porphyria. He had already reported a case (no. 34) of urocoproporphyria without skin lesions.[10] This subject may have had latent porphyria, or alternatively, slight skin lesions such as hyperpigmentation may have been overlooked. Three latent cases were detected by me in 1957, and the urinary findings have been recorded by Mentz and Bersohn.[11] Subsequently, Galambos[12] described the case of an American Negro who excreted both uro- and coproporphyrin in excess in the absence of skin lesions. Gross uroporphyrinuria without skin lesions has been noted in 4 Bantu patients by Eales.[13] While too much reliance should not be placed on the ratios of latent to overt subjects found in the 3 samples surveyed, nevertheless they do draw attention to the comparative frequency of latent cases.

In the light of these findings it is suggested that, in future studies on the incidence of PCT, particular heed be paid to the age structure of the population and associated conditions such as diabetes.

SUMMARY

1. The overall incidence of porphyria cutanea tarda in an adult urban Bantu outpatient population was found to be 1·3%.

2. The incidence is not significantly different in the 2 sexes, but does increase progressively with advancing age.

3. A remarkably high incidence of 4·0% was found in patients attending the diabetic clinic.

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