is now inserting the loop on the 4th day postpartum.

Other sister's own's anomaly in Hirschsprung's disease is from Hirschsprung's disease, consanguinity.

In 3 cases the parents of the studied cases were not available with which to compare the findings of this investigation, so no conclusion can be drawn about this factor in relation to Hirschsprung's disease.

Medical and social workers must be trained in the many facets of family planning, and Government must provide for the availability and distribution of contraceptives to all members of the community.

I don't think this is the place, nor have we the time, to discuss the various methods of contraception. Suffice it to say, that the method for the masses must be acceptable, safe, reasonably effective, cheap, easily distributed and requiring a minimum amount of intelligence and reliability to use with a minimum of patient contact.

All methods have certain disadvantages, but the more knowledge that can be gained about reproductive physiology the more likely is it that we shall arrive at the ideal contraceptive.

Conclusion

The time has come for us to evaluate our position in South Africa. We here, are at the gateway to a vast hinterland peopled by a nation of comparatively low socioeconomic status and education. Illegitimacy is rife but it is beginning to be recognized that family limitations and advantages must eventually lead to a higher standard of living.

Perhaps Professor de Villiers of Cape Town has the answer — he is now inserting the loop on the 4th day postpartum and seeing it into the cervix and his results are distinctly encouraging.

It has taken me approximately 20 minutes to read this paper to you and during that time 2,000 babies have been born.

A GENETICAL STUDY OF HIRSCHSPRUNG'S DISEASE*

CONGENITAL INTESTINAL AGANGLIONOSIS

H. GORDON, B.SC., M.D., M.R.C.P.; MARIE TORRINGTON, M.SOC.SCI., Comprehensive Medicine Group, Department of Medicine; J. H. LOUW, CH.M., F.R.C.S., F.A.C.S. and S. CYWES, M.MED. (SURG.), Department of Surgery, University of Cape Town

This investigation was centred round 71 index cases who had been operated on for Hirschprung's disease at the Red Cross War Memorial Children's Hospital from 1957 till 1966. In each case the diagnosis was confirmed histologically by the absence of autonomic ganglion cells from the wall of the rectum and for a variable but continuous length of gut above the rectum.

Race. Of the 71 index cases, 26 were Whites, 38 Cape Coloured and 7 African. Several extraneous factors influenced the admission of these 71 index cases to this hospital so the data cannot be used for assessing the frequency of the disease in South Africa or for determining whether there is any racial difference in susceptibility to the disease.

Sex. Of the 71 index cases, 55 were males, giving a sex ratio for the whole series of 3:4:1. This is very close to the sex ratios reported in two other investigations from Bremen and London.

Type of lesion. In 53 cases (75%), the lesion was of the 'short segment' type, i.e. it did not extend above the sigmoid colon. In the remaining 18 (25%) a varying length of gut above the sigmoid colon was affected ('long segment' type); in one of these cases the lesion extended into the distal part of the jejunum. In the 'short segment' group, the male:female ratio was 4:9:1; in the 'long segment' group it was just 1:6:1. There was no disproportionate frequency of either type of lesion among the 3 racial groups.

Of the 71 index cases, it was possible to contact the families of 56 directly; these are referred to as the 'studied cases'. Contact could not be established with 15 families who lived far away from Cape Town and whose whereabouts could not be ascertained. Missing names and history of the 'studied cases' only.

Maternal age. The mean age of the mothers at the birth of the affected children was 32.5 years. No data were available for the general population so no comparison between the ages of affected and 'control' fathers could be made.

Birth order. No data from the general population are available with which to compare the findings of this investigation, so no conclusion can be drawn about this factor in relation to Hirschprung's disease.

Maternal health in pregnancy. The retrospective interrogation of the mothers about illnesses, drugs, X-rays, etc. during their pregnancy did not reveal any obviously relevant factor.

Delivery. No undue difficulties at the time of delivery were recalled by the mothers of the studied cases.

Parental consanguinity. In 3 cases the parents of the studied cases were first cousins once removed (coefficient of relationship, r=1/16). For all 56 studied cases, the mean coefficient of relationship, F=0.0016. The presence of this degree of parental consanguinity suggests the possibility of a recessive pattern in the genetics of Hirschprung's disease.

Affected relatives. In only one of the families studied was there histologically confirmed evidence of more than one affected individual. In this family there were 2 affected sisters and a maternal first cousin (male) with Hirschprung's disease. In one of the sisters the lesion was of the long-segment type; in the other sister it was of the extra-long-segment type; and in the cousin it was of the short-segment type. In two other families there was well-documented evidence of an older brother of the index case dying from a disease clinically and radiologically indistinguishable from Hirschprung's disease, but without histological proof. There were no twins in this series.

Associated malformations. Anophthalmos, Down's anomaly, endocardial fibroelastosis, ureteric valves, and mental defect were noted once each among the studied cases. The association with Down's anomaly has often been recorded. If our series is added to 3 others which are adequately documented, the incidence of Down's anomaly in Hirschprung's disease is 8 in 633 cases; in the general population, the incidence of Down's anomaly is about 1 in 600. The association with anophthalmos is interesting. This anomaly may be due to a...
disturbance in the outgrowth and migration of cells from the cranial-cervical portion of the embryonic neural tube; and it is from this region of the neural tube that the ganglion cells of the terminal intestine are believed to originate. It is noteworthy that 2 index cases had first cousins with congenital ptosis and another 2 index cases had first cousins who died with spina bifida; these lesions may also be associated with disturbances of development affecting the embryonic neural tube.

CONCLUSIONS

The findings in this South African series support those from Europe and North America in emphasizing the importance of genetic factors in the aetiology of Hirschsprung's disease. Neither our data nor those from elsewhere are suggestive of either a dominant or an X-linked pattern of inheritance. The occurrence of parental consanguinity and the presence of affected relatives in the pedigrees of a few of our studied cases are consistent with a recessive type of inheritance, but no more definite statement about the genetics of Hirschsprung's disease can yet be made. The tendency of the mothers of our studied cases to be older than in the general population points to environmental influences acting at the genetic level. The association with ocular lesions points to the cranial-cervical portion of the neural tube (rather than the neural crest) as the possible site of the primary embryological defect.

ADDITIONAL FUNDING

Advantage was taken of the facilities of the Ecology of Hypertension Research Project to carry out this investigation. This project is sponsored by the National Institutes of Health of the US Public Health Service (Grant HE 06267). The additional expenses of this investigation were met by a University of Cape Town Staff Research Grant (C.L. Herman Research Fund).

ALLERGENIC PROPERTIES OF SWEAT IN MILIARIA: A NEGATIVE REPORT


In a previous publication it was shown that the earliest histopathological changes in miliaria (prickly heat) resembled those of eczema located around the sweat ducts. These changes precede the obstructive ones which are regarded as the classical features of miliaria; in order to find out whether sweat was acting as an allergen and thus causing miliaria, the following experiments were made. The problem is similar to that of the urticariogenic properties of sweat investigated by other authors, but in this enquiry the delayed, eczematous reaction had to be considered in addition to the immediate, urticarial response.

MATERIAL AND METHODS

Three groups of adult subjects were used in the experiments; they were made up as follows:

1. Eleven miliaria patients of whom 5 presented evidence of the disease at the time of testing.
2. Eight with other skin conditions (5 cement dermatitis, 2 bacterial eczema and 1 primary irritant dermatitis).
3. Two with no skin disease.

Forced sweating was induced in a hot box and when free perspiration was present, usually after half an hour, the back was washed with distilled water, dried and then watched for the reappearance of sweat; this was then collected in test tubes and used for subsequent experiments.

Urticariogenic Properties

Using tuberculin syringes, intradermal injections of 0.1 ml. were made in a total of 9 patients (4 from group 1 and 5 from group 2). The test material consisted of:

1. Unsterilized sweat, from self and other patients.
2. Pasteurized sweat, from self and other patients.
3. Sweat including 0.0001 % of merthiolate solution.
4. Normal saline control.
5. Subject's own sweat untreated.
6. Subject's own sweat pasteurized.
7. Subject's own sweat + merthiolate, 0.0001 %.
8. One other subject's sweat untreated.
9. One other subject's sweat pasteurized.
10. One other subject's sweat + merthiolate, 0.0001 %.

A small square of gauze was dipped in the test solution, applied to the skin and was covered with a 1 sq. in. piece of cellophane and secured with Elastoplast. The patches were removed after 48 hours and read 20 minutes later.

Only 1 sample of sweat gave a strong reaction. The same sweat however, when pasteurized, gave a negative reaction. This sample came from a patient with severe bacterial eczema; his untreated sweat also gave a ++++ patch test reaction in another subject though it gave a negative result when pasteurized. It was therefore concluded that this reaction was due to bacteria or their products and not sweat per se. The saline control tests were also negative but 8 out of 16 cases gave a positive reaction with bichromate patch tests; 5 of these were from group 1, the rest from group 2.

CONCLUSION

None of the 11 miliaria patients showed either an urticarial or eczematous reaction to his own sweat or that of another subject, whether the latter suffered from miliaria or not. The sweat of miliaria subjects produced no reaction in control subjects either by patch test application or by intradermal injection.

SUMMARY

Intradermal and patch testing in miliaria (prickly heat) subjects and controls showed no evidence of allergenic properties in the sweat.

This work was supported by a grant from the Rand Mutual Assurance Company Ltd., Johannesburg.

REFERENCES