DR. J. L. A. PEUTZ
~~REZUIDENHOUT 20~~ L.v.N.O. India 193

DEN HAAG, May the 8th 19.48

Dr. Victor A. McKusick M.D.
The Johns Hopkins Hospital
Baltimore 5, Maryland.

Dear Sir,

In reply to Your letter of April the 13th 1948 concerning the communication published by me in 1921 for the first time: „A most remarkable combined familiar polyposis of the mucous membranes of the trachea intestinalis with those of the naso-pharynx and attended with peculiar pigmentations of skin and mucous membranes" (Nederlandsch Maandschrift voor Geneeskunde, file X [New Series, file 11, Nr. 3, p. 134-146] I can give the following answer.

Indeed the combination of polyposis of the whole trachea intestinalis and of the mucous membranes of the nose-cavity with extensive pigmentations of the skin and the mucous membranes has still sometimes been observed by me among some more relations since my publications in 1921. As to the cases published in 1921 I must observe that in one of these cases moreover polyps were found in the urine-bladder.

As to the pigmentations these were already seen in the mouth-cavity in the beginning second year of live, some years later also on the lips and soon over the whole face and on the neck. Among the members of this family, in so far as I had the opportunity to examine them I found rectal pigmentations earliest at the age of four, probably however they were already there before. The first sort-colaured or dark-brown skin- and lip pigmentations gradually faded in the course of years, and even disappeared for the greater part between the ages of 25 and 30; the spots got at least much smaller, as is also to be seen on the two subjoined photos ([redacted] and [redacted]). The rectal and mouth pigmentations did not undergo this fading and becoming smaller.

Of the relations mentioned in my publication of 1921 in the mean time died, after repeated nose-throat operations and on the intestines [redacted] (1937, ileus), [redacted] (1947, ileus) and [redacted] (1948, ileus). With William already in 1921 a malignant degeneration of some resected intestine-polyps was found by my friend the later Nobelprize-laureate, prof. Landsteiner, then pathologist-anatomist of the R. C. Hospital at the Hague. [redacted] however is still alive (1948). [redacted], [redacted] and [redacted] married; their marriage however remained without children. Our present pathologist-anatomist, Dr. Wijers, found on operation-resection with [redacted] († 1947) and with [redacted] († [redacted] 1948) adeno-carcinomatous degeneration of the resected polyps

From the marriage of [redacted], an extremely serious case with repeatedly recurrent extensive nasal polyps as big as grapes to and fro (cf. enclosed photo), seven children were born, four boys and three girls. Two of these boys and a girl appeared to have again the described syndrome, the eldest [redacted], whose photo I enclose like that of his mother [redacted], a very intelligent boy who already obtained a scholarship, died in 1940 after a serious attack of an ileus not paid much attention to by his family-doctor and to post mortem examination of his gastro-intestinal polyps (proved by rectoscopy and Röntgen) could not be performed, alas!

Besides I got to know another (3rd) sister of the father of the children described in 1921, a [redacted] who died in 1947 of an „abdominal disease". She, too, had anamnestically polyposis of the whole tractus of intestinalis, and skin- and mucous membrane-pigmentations, missed however the nose-polyps present with the others.

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It is of course extremely interesting to trace the further course of life of the children of [redacted] (the eldest, [redacted] has died) in case of a possible marriage, i.e. if in the next generation the syndrome finds progress again.

Since my publication of this very remarkable syndrome (1921) Dr. Cyrax published in the Nederlandsch Tijdschrift about a familiar general polyposis in which however there were no skin- and mucous membranepigmentations

The extremely remarkable syndrome, described for the first time by me in 1921 and further observed and followed by me in the later years up till now, consequently consists of a familiar, hereditary polyposis of the whole tractus intestinalis and of the mucous membranes of the naso-pharynx [and in one of the cases also of the urine-bladder], attended with grotesque pigmentations of the mucous membranes and of the skin, appearing already very early, which pigmentations in so far they are situated on the skin-surface begin to fade towards the age of twenty, and disappear for the greater part. The pigmentations of the mucous membranes of the mouth-cavity and of the rectum, appearing in the first year of life, are however also maintained after twenty.

The inclination for carcinomatous degeneration of a polyadenomatosis, mentioned in literature as a leading thread, did not show clearly here. Only in ~~two cases~~ three of my cases such a degeneration was observed till now: [redacted] (by prof. Landsteiner in 1921) - he is still alive (1948), [redacted] only died in 1947 of ileus, the eliminated operation-piece showed now a malignant degeneration (Dr. Weyers) and [redacted], died in [redacted] 1948 and was only then found to be also a malignant degeneration. (Dr. Weyers)

Prof. Siemens at Leyden with whom I discussed afterwards the syndrome, informed Mairowsky of this who referred to this, after a correspondence with me, in Jadassohn's Handbuch. Mairowsky genealogical chart however does not seem to be quite exact.

Trusting to have been of some service to you in this respect, and kindly requesting you to send some copies, when you proceed to the publication of your cases — of course I take an extraordinary interest in them, I am

Sincerely yours

Dr. J.L.H. Pautz,
Specialist in internal medicine
Director of the department of intern. medicine
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