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CLINICAL CASE

VON RECKLINGHAUSEN'S DISEASE AND CONGENITAL SYPHILIS

Dr. Ironside, at a meeting of the Medical Society for the Study of Venereal Diseases, held on July 9th, 1926, exhibited a girl, Violet B.,* aged ten years, one of two sisters in a family. She was accompanied by her mother. The girl was the subject of von Recklinghausen's disease and congenital syphilis, probably a unique association. This patient's mother acquired syphilis and transmitted it to the two children; both children had been treated intensively at the East Suffolk Hospital by Dr. Fowler Ward, and in the case of the present child the Wassermann reactions in the blood and cerebro-spinal fluid were now negative. The child's facies was suggestive of congenital syphilis.

The von Recklinghausen's disease was also inherited from the maternal side. It could be traced through four generations. Dr. Ironside had seen the child's maternal grandfather, who had typical sessile and pedunculated mauve-coloured and coffee-coloured neuromata in the skin. The child's mother (demonstrated) had coffee-coloured patches of von Recklinghausen's disease on the neck and the typical pedunculated neuromata together with a plexiform neuroma on the scalp. The child shown and her sister had the disease in a more abortive form and showed coffee-coloured patches on the abdomen and back, and only three nodules (sessile) in the skin. There were no mauve-coloured nodules. The disease seemed to lessen in intensity in its cutaneous manifestations in succeeding child-ranks.

The child exhibited was obese, the skiagram of the skull showed an enlarged pituitary fossa, the sugar tolerance was increased, and she had papilloedema subsiding into secondary optic atrophy.

The patient's sister, whom Dr. Ironside had seen at

* Shown by the kindness of Dr. W. J. Adie.
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Ipswich two years ago, had since died, and was found at autopsy to have a supra-pituitary cyst.

The question arose as to how much of Violet B.'s symptoms was due to the von Recklinghausen's disease, and how much to syphilis. A lumbar puncture had been made and the fluid was found to be normal in all respects. The association of von Recklinghausen's disease with cystic formation in the long bones was mentioned. Pituitary symptoms had been noted in cases of von Recklinghausen's disease.* Tumours of nervous tissue might occur in the peripheral or central or in the sympathetic nervous system.

The President said the sessile neuromata were more tender, usually, than the pedunculated ones. He asked whether Dr. Ironside agreed that von Recklinghausen's disease was a form of naevus; that was the general view. In a case he had fifteen years ago, with the late Sir Malcolm Morris, there were six tumours, which were excised, a different anaesthetic being used with each one, as they were so tender. The sessile ones consisted of a translucent gelatinous mass.

Dr. Ironside replied that he thought von Recklinghausen's disease was an hereditary and congenital disease with nervous, cutaneous, osseous and endocrine manifestations. The tumours of nervous tissue were, he thought, a benign hypertrophy of parenchymatous or interstitial tissue.

The President said that the disease was commonly familial.