Kallmann's Syndrome

'A rose by any other name would smell as sweet.' (Shakespeare)

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Kallmann's Syndrome (KS), or hereditary hypogonadal anosmia, is a rare genetic disorder characterised by delayed or absent puberty and anosmia. It is named after Franz Kallmann, but he was certainly not the first doctor to describe the condition. At least two others had pipped him to the post. In 1856 the Spanish anatomist, Aureliano Maestre de San Juan, reported a patient with total absence of the olfactory nerves, a microphallus and testicular atrophy [1]. Then, in 1861, Herschel of Vienna described a patient with 'testes which were the size of beans,' a 'feminine larynx (sic)' and anosmia [2].

Franz Josef Kallmann (1897 –1965) who described the disorder in 1944 [3] was born in Silesia, Germany, which later became a part of Poland. He was the son of a surgeon and general practitioner. In Munich he worked with the famous Swiss-born psychiatrist and geneticist, Ernst Rüdin (a man whom Hitler later called the 'pathfinder in the field of hereditary hygiene'). Kallmann was Jewish and fled Germany in 1936, but in 1935 he gave a speech while still in Germany, in which he advocated the examination of relatives of schizophrenics to see whether or not they were 'non-affected carriers' of the supposed recessive schizophrenia gene. If they proved positive, the plan was to sterilise them [4].

Some cases of KS are obvious at birth because of the extremely small penis (microphallus). In the past, when all UK state schools had a school doctor, this and undescended or rudimentary testicles would certainly have been detected at a routine school medical examination. The syndrome occurs in both sexes, but is more common in males. There is no growth spurt. Also the boy's voice does not break and there is a lack of beard growth. In girls, primary amenorrhoea is the most usual presentation. Interestingly in most cases, patients had not realised they had no sense of smell.

Aureliano Maestre de San Juan.
Source: Image bank of the Royal National Academy of Medicine, Spain.

Franz Kallmann, 1950.

There are quite a few other associated abnormalities, among the most fascinating of which is a neurological sign called bimanual synkinesis. This is when the movements of the dominant hand are mirrored by the non-dominant one. This rare symptom can make it difficult to perform tasks that require the hands to move independently.

One man cashed in on his Kallmann's syndrome. 'Little Jimmy Scott' became a well-known jazz vocalist in the 1940s, famed for his high boy-soprano voice [5]. He initially only grew to a height of four feet 11 inches. Then, when he was 37 years old, he grew another eight inches and went on to live to his 89th year (although strenuously denied, hormone therapy was strongly suspected).

Eponymous names (particularly those associated with a eugenicist) are currently controversial and some would suggest that Kallmann's Syndrome should now be called 'congenital anosmic idiopathic hypogonadotropic hypogonadism'. I leave my readers to make their own decision here.

References

1. Maestre De San Juan A. Teratologia; Falta total de los nervios olfactorios con anosmia en un individuo en quien existia una atrofia congenita de los testiculos y miembro viril. *El Siglo Medico* 1856;**131**:211.
2. Mackenzie, JN. The Physiological and Pathological Relations between the Nose and the Sexual Apparatus of Man. *Bull Johns Hopkins Hosp* 1898;**IX(82)**.
3. Kallmann FJ, Schonfeld WA, Barrera SE. The Genetic Aspects of Primary Eunuchoidism. *Am J Ment Defic* 1944;**48**:203–36.
4. Joseph, J. *The Gene Illusion: Genetic Research in Psychiatry and Psychology under the Microscope.* New York, USA; Algora; 2004.
5. For a complete discussion of the history of Kallmann's Syndrome see Chapter 8 in Young JR. *Sex and the Nose.* Amsterdam, The Netherlands; Kugler; 2020.