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Kallmann syndrome: A failure of GnRH neuronal migration

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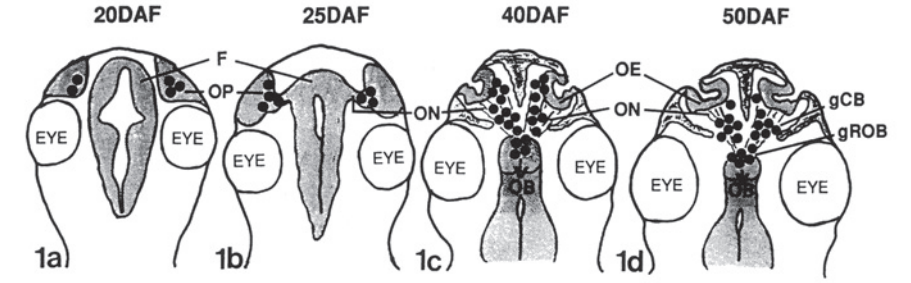

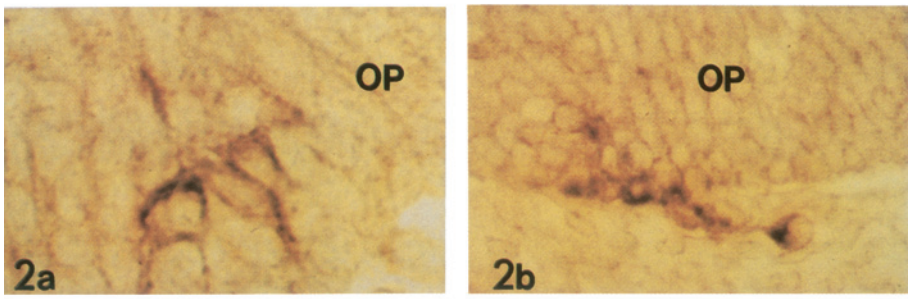

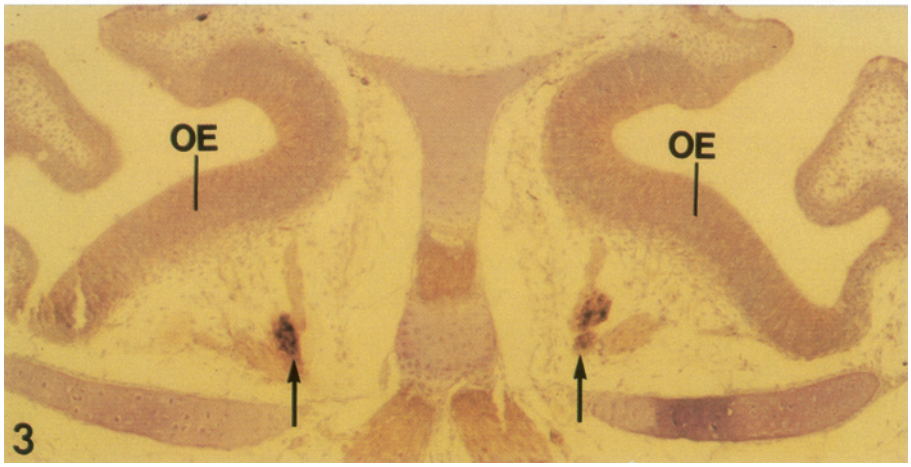

Gonadotropin-releasing hormone（GnRH）neuronal migration from the olfactory placodes to the basal forebrain is a vertebrate-wide phenomenon seen in species ranging from fish to humans. The failure of these neurons to migrate leads to Kallmann syndrome（hypogonadotropic hypogonadism coupled with anosmia).

Fig. 1 a–d　Embryonic fish（20–50 days after fertilization, DAF）. Camera lucida drawings show the migratory route of GnRH neurons from the olfactory placodes（OP）to the basal forebrain（F）. Olfactory epithelium(OE).

Fig. 2 a, b　GnRH-immunoreactive neurons seen in the olfactory placodes on(a)embryonic day 25 and(b)day 30.

Fig. 3–4　GnRH neurons in a ganglia along the olfactory nerve(ON)at day 100.

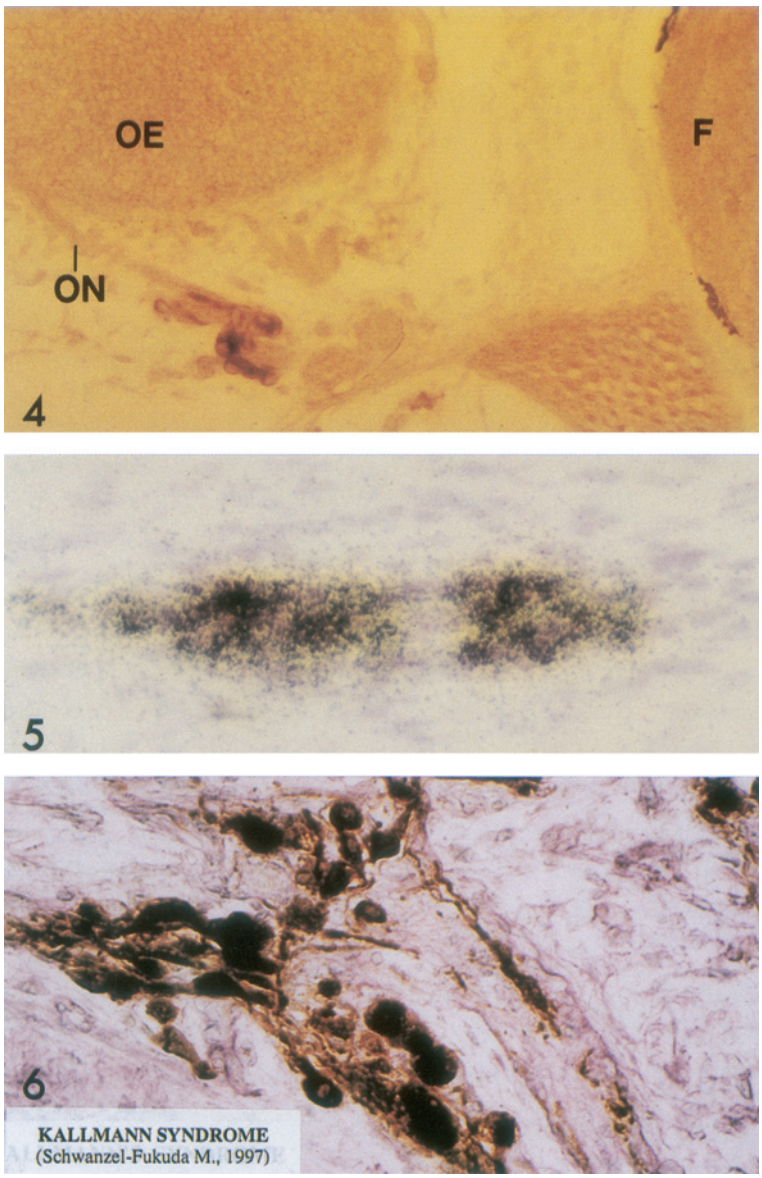

Fig. 5 Neurons expressing GnRH mRNA hybridization signals along the olfactory nerve.

Fig. 6 Nasal region of a 19 week-old human fetus with Kallmann syndrome. A cluster of GnRH neurons seen " dammed up "at the top of the olfactory apparatus. No GnRH immunoreactivity was found in the brain, and both olfactory bulbs were absent (Schwanzel-Fukuda, 1997).

解説：視床下部の神経細胞により産生され，下垂体前葉の分泌を調節する物質を放出ホルモンとよぶ．性腺刺激ホルモンの放出を起す性腺刺激ホルモン放出ホルモン（GnRH）を産生するニューロンは，嗅上皮の原基である嗅板 olfactory placode に発症して，脳に入り込み，視床下部に移動するという特異な個体発生を経過する．嗅板から脳への移行が阻止されたために生じた病態が Kallmann syndrome で，嗅覚の欠損と中枢性の性腺機能低下を呈する．

Reference

Schwanzel-Fukuda M: The origin and migration of LHRH neurons in mammals: A comparison between species including human. In: Parhar I.S. and Sakuma Y.(eds). GnRH Neurons, Gene to Behavior. 1997; pp 221-242. Brain Shuppan Publishers, Tokyo.