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Pathogenesis of an ectopic posterior lobe in patients of short stature with growth hormone deficiency.

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LETTERS

Coil Embolization of Posterior Inferior Cerebellar Artery Isolated Aneurysms

I read with great interest the article by Tikkakoski et al (1) describing their treatment of an isolated dissecting aneurysm of the left posterior inferior cerebellar artery with Guglielmi detachable coils (GDCs). I would like to draw the authors' attention to two additional articles relating to this subject. One article published in the *Journal of Neurosurgery* was a case report of an isolated posterior inferior cerebellar artery treated with standard platinum microembolization coils. The lateral medullary syndrome was fortunately avoided in this patient (2). Another case that we previously reported was a pseudoaneurysm of the posterior inferior cerebellar artery, caused by a head and neck tumor biopsy, which was treated by proximal occlusion of the posterior inferior cerebellar artery using standard platinum microembolization coils (3). I hope these two additional articles will show that in some situations, isolated aneurysms involving the posterior inferior cerebellar artery need not be occluded with GDCs, but can be treated with more conventional coil devices. However, the authors have achieved an excellent result with GDCs.

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Reply

We appreciate the interest in our case report expressed by Dr Teitelbaum. Dissecting aneurysms of the posterior circulation are rare. Only few reports on their endovascular treatment, with conventional coils, balloons, or both, exist. Advantages of the GDC technique are the predictability and controllability of the coil before detachment. Disadvantage is the cost. To our knowledge, our 34-year-old woman with the progressive dissecting aneurysm of the posterior inferior cerebellar artery was treated (June 1995) with a GDC and was doing well in May 1997.

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Pathogenesis of an Ectopic Posterior Lobe in Patients of Short Stature with Growth Hormone Deficiency

Magnetic resonance (MR) imaging has shown characteristic findings in patients of short stature with growth hormone deficiency: pituitary stalk disruption and an ectopic posterior lobe above the stump of the disrupted stalk (1-5). However, there has been a controversy concerning pathogenesis of this entity, and two hypotheses have been proposed. One is that the ectopic posterior lobe is formed above the stump of the stalk, which is transected mechanically during breech presentation or head trauma (1-3). The other is the maldevelopment hypothesis (4, 5).

The maldevelopment hypothesis is based on the fusion concept in an embryonal development of the pituitary gland (Fig 1)

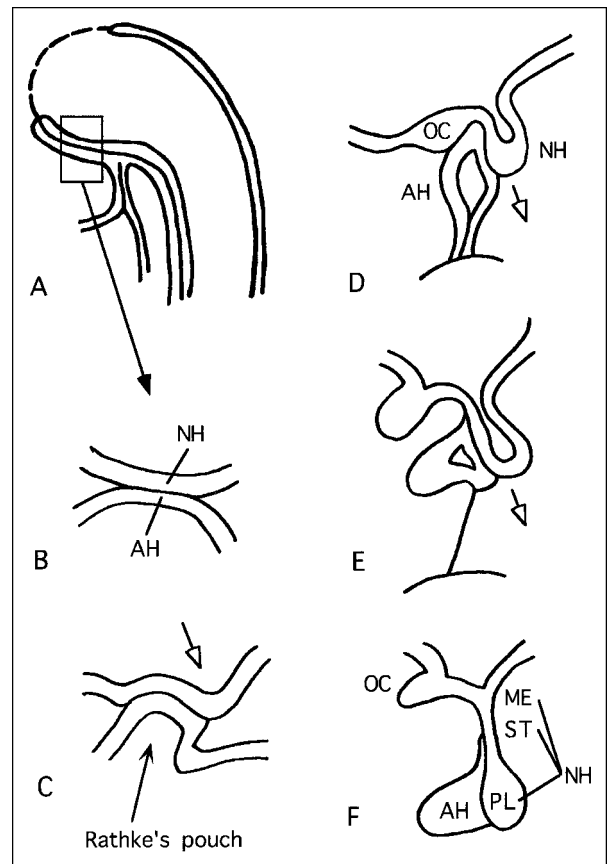


FIG 1. The fusion concept in the embryonal development of the pituitary gland and the maldevelopment hypothesis in the pathogenesis of the ectopic posterior lobe, based on Kelly et al (4).

A and B, The fusion concept in organogenesis of the pituitary gland that is widely believed but apparently incorrect (compare with the true embryology in Figure 2). The primordia of the adenohypophysis and neurohypophysis are separated at the beginning. Both structures fuse to form the pituitary gland during development.

C, The maldevelopment hypothesis. The ectopic posterior lobe is caused by incomplete downward growth of the neurohypophysis.

AH indicates adenohypophysis; NH, neurohypophysis; OC, optic chiasm; EPL, ectopic posterior lobe; ME, median eminence; ST, stalk; and PL, posterior lobe.

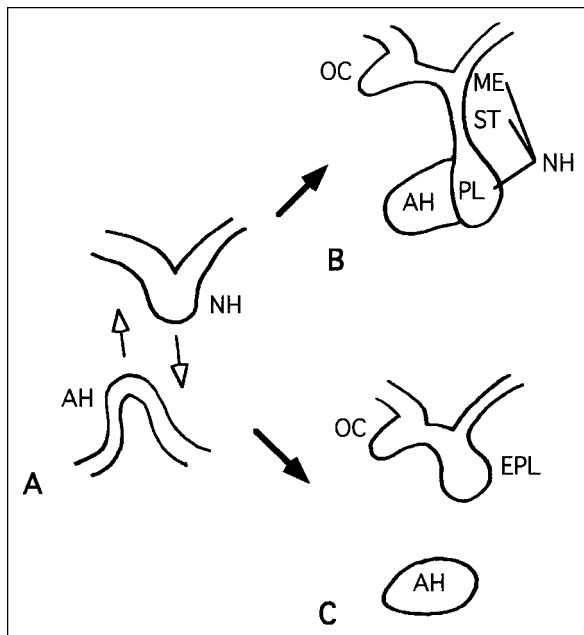


FIG 2. True embryonal development of the pituitary gland, based on Romeis (6) and O'Rahilly and Müller (7).

A, Median section at 4½ weeks.

A and B, The primordia of the adenohypophysis and neurohypophysis form a single structure from the beginning.

C, At 6 weeks, an evagination of the neurohypophysis (arrow) and Rathke's pouch are formed.

D-F, The neurohypophysis and adenohypophysis grow downward together to form the pituitary stalk and gland. The maldevelopment hypothesis (Fig 1C) is contradictory to the true embryonal development of the pituitary gland because the adenohypophysis and neurohypophysis are not separated throughout embryonal development.

AH indicates adenohypophysis; NH, neurohypophysis; OC, optic chiasm; ME, median eminence; ST, stalk; and PL, posterior lobe.

(4, 5). The anterior (adenohypophysis) and posterior (neurohypophysis) lobes are separated in the early embryo and fuse into the whole pituitary gland (Fig 1 A and B). The anterior lobe is derived from ectodermal development of the stomodaeum. The neurohypophysis develops as a downward evagination of the neuroectoderm of the floor of forebrain. The maldevelopment hypothesis suggests that the pathogenesis of the ectopic posterior lobe is incomplete downward growth of the neurohypophysis (Fig 1C).

The fusion concept is widely held among clinicians. Personally, I believe it is incorrect. In fact, the fusion of the adenohypophysis and neurohypophysis never occur in the embryonic brain (6-8) (Fig 2). Both structures are in apposition from the beginning and do not have to grow toward each other (8). Primordia of the adenohypophysis and neurohypophysis form a single structure in the early embryo and extend downward together to form the pituitary stalk and gland (Fig 2). This is a long-established concept among embryologists (6), mistranslated to clinicians as the fusion concept.

The maldevelopment hypothesis is contradictory to the true embryology of the pituitary gland. I suggest that the pathogenesis of the ectopic posterior lobe is the mechanical transection of the pituitary stalk during breech presentation or head trauma.

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Reply

In 1987, Dr Fujisawa described ten patients with congenital growth hormone deficiency, a previously unreported feature defined as "ectopic neurohypophysis" (1). He correctly identified the ectopic bright spot as the neurohypophysis, having previously understood and reported that the intrasellar bright spot was the neurohypophysis itself (2). His interpretation for the bizarre ectopic position of the bright spot was that a traumatic delivery had produced a transection of the pituitary stalk and hence a proximal concentration of the components responsible for the hyperintensity of the posterior pituitary. This interpretation was probably attributable to the fact that in his original series almost all the patients had had a breech presentation (which can be a cause for traumatic delivery). In that same year, we had the opportunity to study some similar patients (3), and by 1992 studied more than 100 (4). By carefully analyzing many factors in our series, gradually we found some pieces of evidence that argued against the "traumatic delivery" hypothesis: (a) 54% of patients with posterior pituitary ectopia had a normal presentation (cephalic) and a normal delivery, (b) 14% of patients had a cesarean delivery, and (c) in the remaining 32% of patients with breech presentation and delivery, no traumatic birth was reported (a breech presentation and delivery does not necessarily imply a traumatic event). We proposed a new hypothesis based on maldevelopmental midline embryologic structures in the area of the pituitary gland and hypothalamus. This hypothesis was based on modern embryologic concepts, partly rediscovered now by Dr Fujisawa.

Some experimental works of cellular embryology in chimeras (5) demonstrate that hormone-secreting cells of the adenohypophysis are more than "close" to the neurohypophysis and are actually part of the neural tube itself, only secondarily migrating to the stromal part of the adenohypophysis that is a contiguous, well-identified part (6). In other words, we never stated that the cause of posterior pituitary ectopia is an incomplete downward growth of the neurohypophysis; we think of a more complex defective induction of primitive mediobasal structures of the brain, possibly on a genetic basis. Some examples of gene malfunction as a cause of hypoplastic pituitary in dwarf mice have been reported (7).

We approached the question of why these patients have such a high percentage of breech presentation. The fetal contribution to its final position in utero in general is still unclear; congenital muscular dystrophies, hypotonic syndromes, and some severe malformations of the nervous system are associated with breech presentation. Breech presentation is exceedingly high (up to 50%) in Prader Willi syndrome (8); these patients present with diffuse hypotonia that could justify an abnormal presentation but also have a definite hypothalamic dysfunction. An abnormal hypothalamus could not respond properly to hormones that could play a crucial role in timing the correct position of the fetus. Can we suggest that in pituitary dwarfs breech delivery is a consequence, and not a cause, of an hypothalamic-pituitary abnormality?

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Association of Posterior Fossa Dermoid Cyst and Klippel-Feil Syndrome

A 40-year-old woman presented to our unit with a 9-month history of occipital headache, exacerbated by coughing. Three weeks before presentation, right-sided facial numbness and paroxysmal weakness of both legs developed, causing her to fall repeatedly, but without loss of consciousness.

On examination, her gait was ataxic and there was upper-limb hyperreflexia. Right-sided sensory deficit of the C-2 and trigeminal dermatomes was shown. There was absence of the right corneal reflex and impairment of the right gag reflex.

Plain radiographs showed the typical Klippel-Feil fusion of cervical vertebrae (Fig 3A). T1-weighted coronal MR images showed a high-signal mass extending from the brain stem up to the tentorium cerebelli (Fig 3B). Sagittal T2-weighted images showed that the same mass had medium signal, with focal areas of low signal in keeping with calcification.

The classic triad of short neck, low posterior hairline, and limitation of neck movement is seen in approximately 52% of

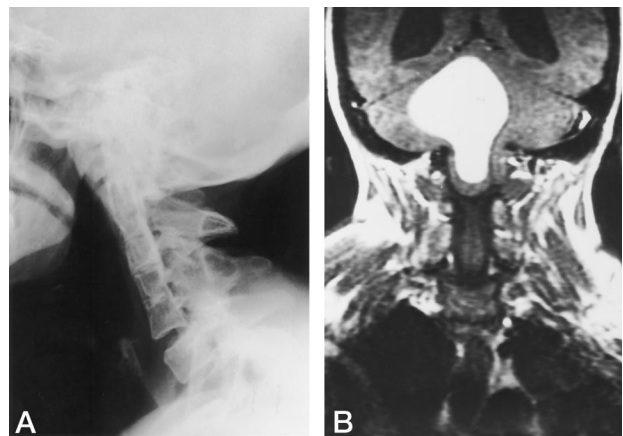


FIG 3. A, Lateral cervical spine radiograph shows the fusion of cervical vertebrae, pituitary fossa erosion secondary to chronically raised intracranial pressure, and calcification projected over the posterior fossa.

B, Coronal T1-weighted MR image (300/16 [repetition time/echo time]) shows the high-signal mass extending from brain stem to tentorium cerebelli, associated with dilatation of the lateral ventricles.

patients with the Klippel-Feil abnormality (1). The syndrome is associated with a range of abnormalities affecting other organ systems, including the central nervous system. Documented associations include diastematomyelia, syringomyelia, and agenesis of the corpus callosum (2), and we are aware of other reports (2-4) of associated posterior fossa dermoid cyst. One of these cases presented with meningism after rupture of the dermoid cyst (2).

Failure of formation and rearrangement of the segmental cervical sclerotomes leads to the Klippel-Feil abnormality. A related, adjacent failure of cleavage of epithelial ectoderm from neuroectoderm may explain an associated dermoid cyst. If a posterior fossa mass is seen on MR or computed tomography in patients with Klippel-Feil syndrome, dermoid cyst should be considered.

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