

The **next generation** GBCA
from Guerbet is here

Explore new possibilities >

Guerbet | 

© Guerbet 2024 GUOB220151-A

AJNR

Duplication of the pituitary gland as shown by MR.

B D Ryals, D C Brown and S W Levin

AJNR Am J Neuroradiol 1993, 14 (1) 137-139

<http://www.ajnr.org/content/14/1/137>

This information is current as
of September 23, 2024.

Duplication of the Pituitary Gland as Shown by MR

Brian D. Ryals,^{1,5,7} Douglas C. Brown,^{2,4} and Sondra W. Levin^{3,6}

Summary: Duplication of the pituitary gland is a rare malformation. The authors describe a patient with features of the median cleft face syndrome (hypertelorism, V-shaped hairline, and developmental delay) who exhibited duplication of the pituitary gland and diffuse thickening of the hypothalamus (hypothalamic hamartoma) as shown by MR. The embryology of the developing pituitary gland and pathogenesis of pituitary duplication are discussed.

Index terms: Pituitary gland, abnormalities and anomalies; Pituitary gland, magnetic resonance

Duplication of the pituitary gland is a rare malformation. Previous reports have noted its association with partial twinning and the median cleft face syndrome (1–5). We report a case where duplication of the pituitary was shown by magnetic resonance (MR) in a patient with mild median cleft face syndrome.

Case Report

The patient was referred for neurologic evaluation at 34 months of age because of developmental delay and atypical facial features. After an uncomplicated pregnancy, he was delivered by cesarean section for early fetal distress with Apgar scores of 7 and 9. He was noted to have mildly dysmorphic facial features, but no other abnormalities on his initial examination. At 6 months of age he was noted to be somewhat hypotonic. He sat unsupported at 10 months, walked at 16 months, and was slow in reaching all milestones. Bayley Scales of Infant Development showed Mental Development Index (developmental age divided by chronologic age multiplied by 100, an index similar but not identical to intelligence quotient) of 61 at 27 months (6). Family history was not revealing.

Physical examination revealed a pleasant and active child. Head circumference was 52 cm (90th percentile for

age), height 95 cm (50th percentile for age), and weight 13.1 kg (25th percentile for age). He had hypertelorism with interpupillary distance of 5.5 cm (>97th percentile for age), a V-shaped frontal hairline, and mild micrognathia; the rest of the physical exam was unremarkable. Complete blood count, electrolytes, glucose, liver enzymes, serum amino acids, thyroid functions, chromosomes, AM cortisol, and somatomedin-C were all normal.

MR of the head revealed hypertelorism as well as thickening of the hypothalamus with normal signal intensity in all sequences. Duplication of the pituitary gland was suspected on the initial routine MR and confirmed with a dedicated sella MR without and with Gd-DTPA infusion. (Figs. 1A–1D). Our diagnosis is presumptive since it is based solely on imaging and there is no surgical or pathologic confirmation.

Discussion

Pituitary duplication is a rare malformation. We found five previous reports in the pathologic literature. The first three reports were seen in association with partial twinning (1–3). More recently, it was reported in association with the median cleft face syndrome (4) and in a newborn who was exposed to meclizine during pregnancy and had multiple congenital defects (5).

Our case appears to represent a mild form of the median cleft face syndrome as opposed to partial twinning. The median cleft face syndrome, which is also known as frontonasal dysplasia, consists of hypertelorism, a low V-shaped frontal hairline, broad nasal root, and varying degrees of midfacial clefting to include cleft nose, lip, palate, and cranium bifidum occultum frontale (7–9). It may be associated with microphthalmia, primary telecanthus, and interhemispheric lipoma (7, 8).

Received August 27, 1991; revision requested December 9, 1991; revision received March 13, 1992 and accepted March 18, 1992

The opinions stated are those of the authors and should not be construed as official or representing the opinions of the Army or Department of Defense.

¹ Child Neurology Service, Walter Reed Army Medical Center (WRAMC), Washington DC 20307-5001.

² Department of Diagnostic Radiology, WRAMC, Washington DC 20307-5001.

³ Department of Pediatrics, WRAMC, Washington DC 20307-5001.

⁴ Department of Radiology and Nuclear Medicine, Uniformed Services University of the Health Sciences (USUHS), Bethesda MD 20814-5001.

⁵ Department of Neurology, USUHS, Bethesda MD 20814-4799.

⁶ Department of Pediatrics, USUHS, Bethesda MD 20814-4799.

⁷ Department of Neurology, Fitzsimmons Army Medical Center (FAMC), Aurora, CO 80015. Address reprint requests to Brian D Ryals, MD.

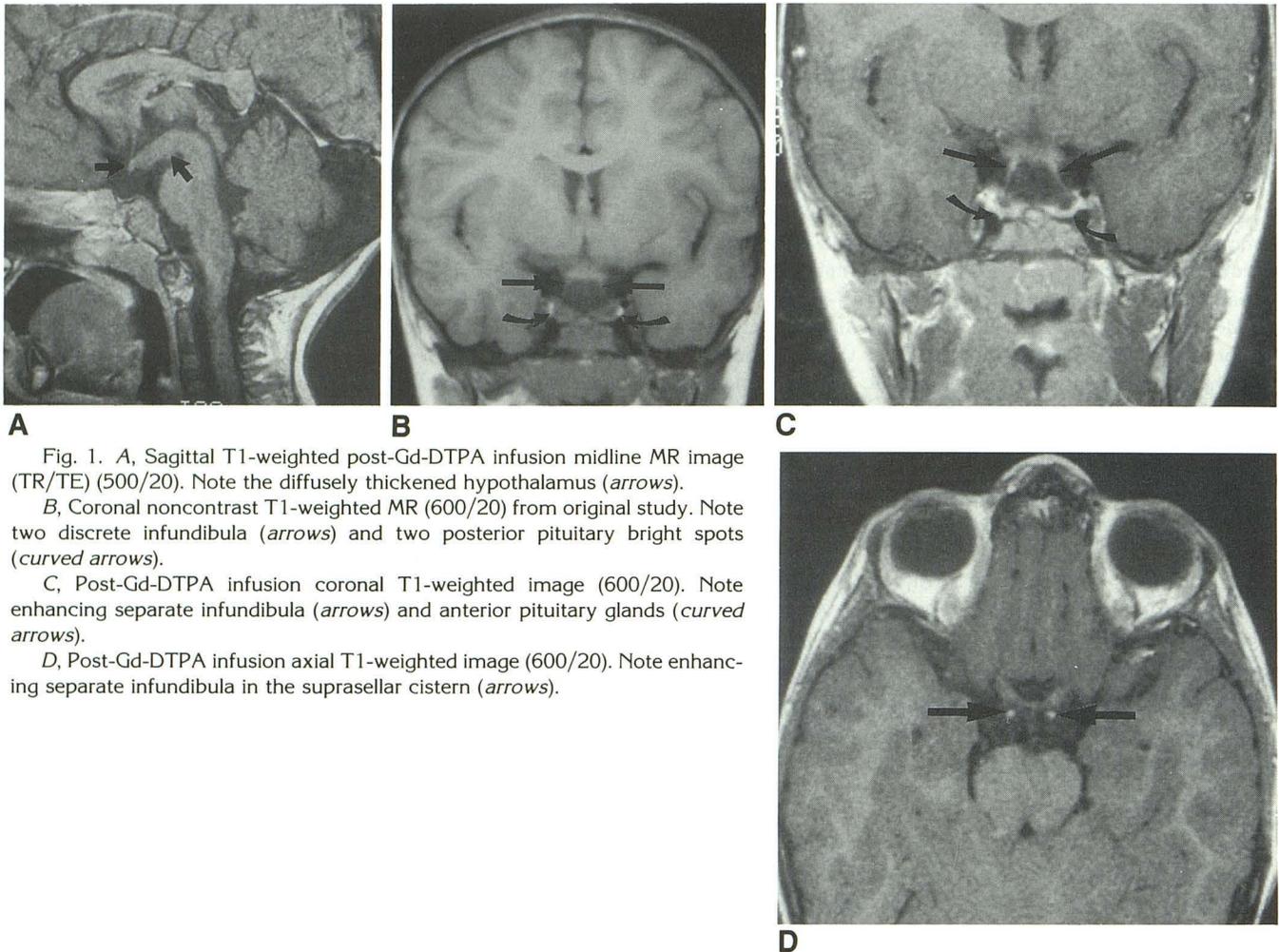


Fig. 1. A, Sagittal T1-weighted post-Gd-DTPA infusion midline MR image (TR/TE) (500/20). Note the diffusely thickened hypothalamus (*arrows*).

B, Coronal noncontrast T1-weighted MR (600/20) from original study. Note two discrete infundibula (*arrows*) and two posterior pituitary bright spots (*curved arrows*).

C, Post-Gd-DTPA infusion coronal T1-weighted image (600/20). Note enhancing separate infundibula (*arrows*) and anterior pituitary glands (*curved arrows*).

D, Post-Gd-DTPA infusion axial T1-weighted image (600/20). Note enhancing separate infundibula in the suprasellar cistern (*arrows*).

Hypertelorism is the only obligatory finding (7, 8). A single report in the pathologic literature described duplication of the pituitary in a neonate with the median cleft face syndrome (4). This child also had agenesis of the corpus callosum, widely separated olfactory nerves, and a thickened hypothalamus of hyperplastic gray matter or a "hypothalamic hamartoma" that laterally displaced the mammillary bodies (4). In our case, we did not appreciate any wide separation of the olfactory nerves and the corpus callosum was intact. Otherwise, our case was strikingly similar to the case described by Hori (4), with a diffusely thickened hypothalamus laterally displacing the mammillary bodies and complete duplication of the infundibulum and both anterior and posterior lobes of the pituitary gland.

Embryologically, the pituitary gland is derived from two ectodermal primordia that unite to form the composite structure (4, 10). During the fourth week of gestation, Rathke's pouch projects dorsally from the roof of the stomodeum (primitive mouth cavity) and grows toward the brain. By

the fifth week, it comes into contact with the infundibulum, a ventral downgrowth of the diencephalon. Rathke's pouch is divided into an upper and lower portion by the primordium of the sphenoid bone. The pharyngeal portion usually degenerates and disappears. Pituitary duplication in partial twinning is thought to result from duplication of the prochordal plate and anterior end of the notochordal process leading to duplication of the entire stomatodeal region to include the pituitary gland (2). The pathogenesis of pituitary gland duplication in the median cleft face syndrome is not clear. It has been suggested that both the developing adenohypophysis from Rathke's pouch and developing neurohypophysis from the diencephalon are completely split related to the median clefting with subsequent development of two separate pituitary glands and infundibula (4). The diffusely thickened hypothalamus (hypothalamic hamartoma) may be secondary to the development of the two separate pituitary glands (4).

In summary, duplication of the pituitary is a rare malformation that may be seen in association with the median cleft face syndrome. Pituitary duplication should be added to the list of abnormalities searched for in children suspected of having midline malformations on MR.

References

1. Ahlfeld F. *Die Missbildungen des Menschen*. Grunow: Leipzig, 1880: 72-95
2. Morton WRM. Duplication of the pituitary and stomatodeal structures in a 38-week male infant. *Arch Dis Child* 1957;32:135-141
3. Warkany J. *Congenital malformations*. Chicago: Year Book, 1975: 419-456
4. Hori A. A brain with two hypophyses in the median cleft face syndrome. *Acta Neuropathol* 1983;59:150-154
5. Tagliavini F, Pilleri G. Mammillo-hypophyseal duplication. *Acta Neuropathol* 1986;69:38-44
6. Louick D, Boland TB: Psychologic tests: a guide for pediatricians. *Pediatr Ann* 1978;7:849
7. Naidich TP, Osborn RE, Bauer B, Naidich MJ. Median cleft face syndrome: MR and CT data from 11 children. *J Comput Assist Tomogr* 1988;12:57-64
8. DeMyer W. The median cleft face syndrome. *Neurology* 1967;17: 961-971
9. Sedano HO, Cohen MM, Jirasek J, Gorlin RJ. Frontonasal dysplasia. *J Pediatr* 1970;76:906-913
10. Moore KL. *The developing human: clinically oriented embryology*. Philadelphia: Saunders, 1988:386-388