Pituitary Gland Duplication and Triplication

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Duplication of the pituitary gland is a rare malformation. It can be complete, involving both the pituitary gland and stalk, or incomplete, affecting the adenohypophysis or the pituitary stalk. Exceptionally the malformation consists in a triplication of the pituitary gland and stalk. The suggested pathogenesis is duplication of the prochordal plate and rostral end of the notochord during early embryologic development. When associated with severe abnormalities, this malformation often results in death within the neonatal period. In other cases, presenting signs are delayed puberty or precocious puberty. It is commonly associated with various midline craniofacial malformations such as hypertelorism, cleft palate, tongue anomalies, choanal atresia, pharyngeal mass protruding in the oral cavity, and persistence of the craniopharyngeal canal. Intracerebral anomalies include thickening of hypothalamus caused by tubomamillary fusion, corpus callosum agenesis, thalamic fusion resulting from mass intermedia agenesis, posterior cranial fossa malformations, olfactory nerve anomalies, circle of Willis anomalies, microcephaly, neuronal migration disorders, hydrocephalus, and spinal abnormalities. Incomplete duplication is considered a less severe malformation, usually not associated with other major anomalies and allowing patients to reach adulthood. Asymptomatic forms have been reported.

In complete duplication, the most constant sign on MRI is a midline elongated hypothalamic mass along the floor of the third ventricle, extending from the optic chiasm to the interpeduncular fossa. This characteristic sign is due to tubomamillary fusion, which is best seen on the midline sagittal image. Fusion of tuber cinereum and mammillary bodies is histologically consistent with an arrest of lateral migration in cells that would normally form the hypothalamic nuclei. This thickening of the hypothalamus leads one to perform thin-section images of the sellar region, which will show two infundibular recesses, two infundibular stalks, and two pituitary stalks extending to two small pituitary glands within two separated pituitary fossa. This is best demonstrated on coronal images (Fig. 57.1). A screening for all the aforementioned intracerebral anomalies should be performed on MR. MRA is recommended for the diagnosis of vascular anomalies, such as basilar artery fenestration. Cranial anomalies such as skull-base defects are better delineated on CT. Differential diagnosis of a mass of the floor of the third ventricle includes hypothalamic hamartoma and hypothalamic glioma. In cases of non-visualization of the pituitary gland or stalk on the midline sagittal MR image, pituitary gland agenesis or hypoplasia and transected pituitary stalk can be distinguished from

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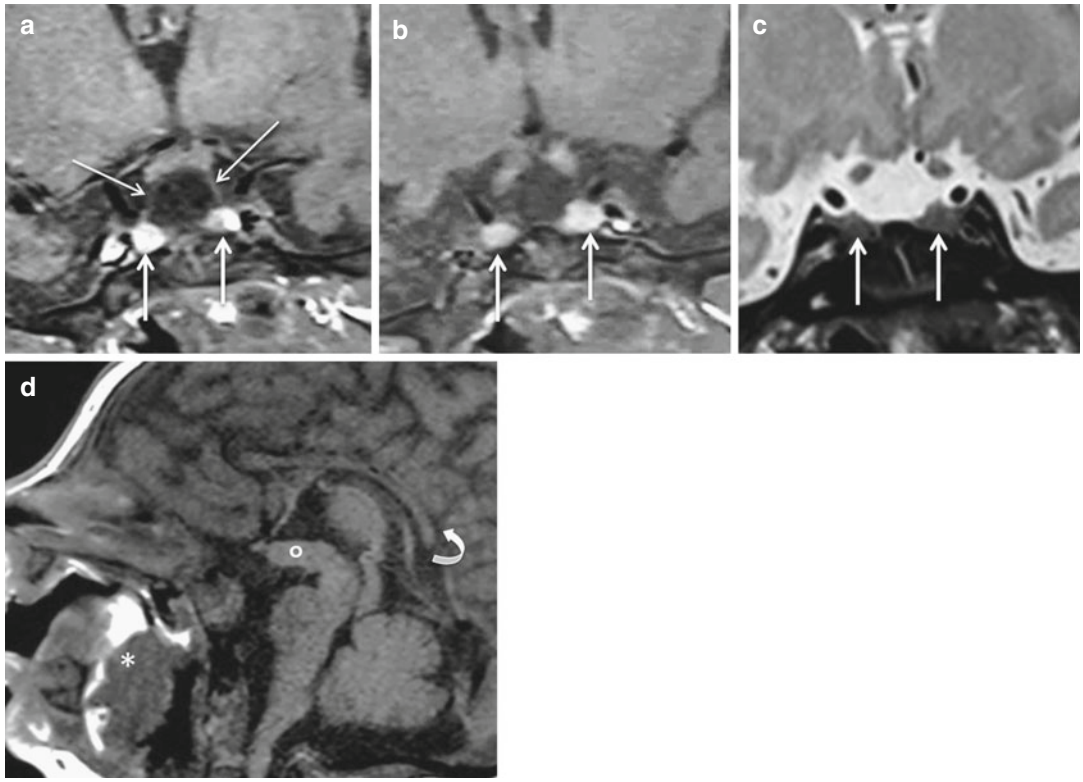

Fig. 57.1 7-day-old boy with mild facial dysmorphism. Duplicated pituitary gland and stalk; mass of the oral cavity. (**a**–**b**) Coronal T1WI and (**c**) T2WI: two pituitary stalks (*long arrows*) and two separate bright pituitary glands (*thick arrows*) within duplicated sella. (**d**) Midline sagittal T1WI: thickening of the floor of the third ventricle resulting from tubomamillary fusion (*circle*). Heterogeneous protruding mass in the oral cavity with fatty component (*asterisk*), the splenium of corpus callosum is thin (*curved arrow*)

complete pituitary duplication by findings in the coronal plane.

Unlike complete pituitary duplication, hypothalamus thickening is absent in incomplete duplication. In incomplete duplication that involves the pituitary stalk, there is a defect of the anterior aspect of the third ventricle floor, enlarging the infundibular recess. Two pituitary stalks, each arising from the floor of the third ventricle, are depicted. Adenohypophysis and sella are single and the posterior pituitary is normal. Anterior third ventricle defect may also suggest the diagnosis of meningocele. The best argument in favor of pituitary stalk duplication is absence of mass effect. Incomplete duplication involving the adenohypophysis is seen on MRI as a linear hypointensity on all sequences, splitting in two the adenohypophysis.

Triplication of the pituitary gland and stalk is exceptional. It can be incomplete with three separate pituitary stalks and a single pituitary gland (Fig. 57.2), or complete with two lateral pituitary glands and a midline gland, each with an independent stalk (Fig. 57.3).

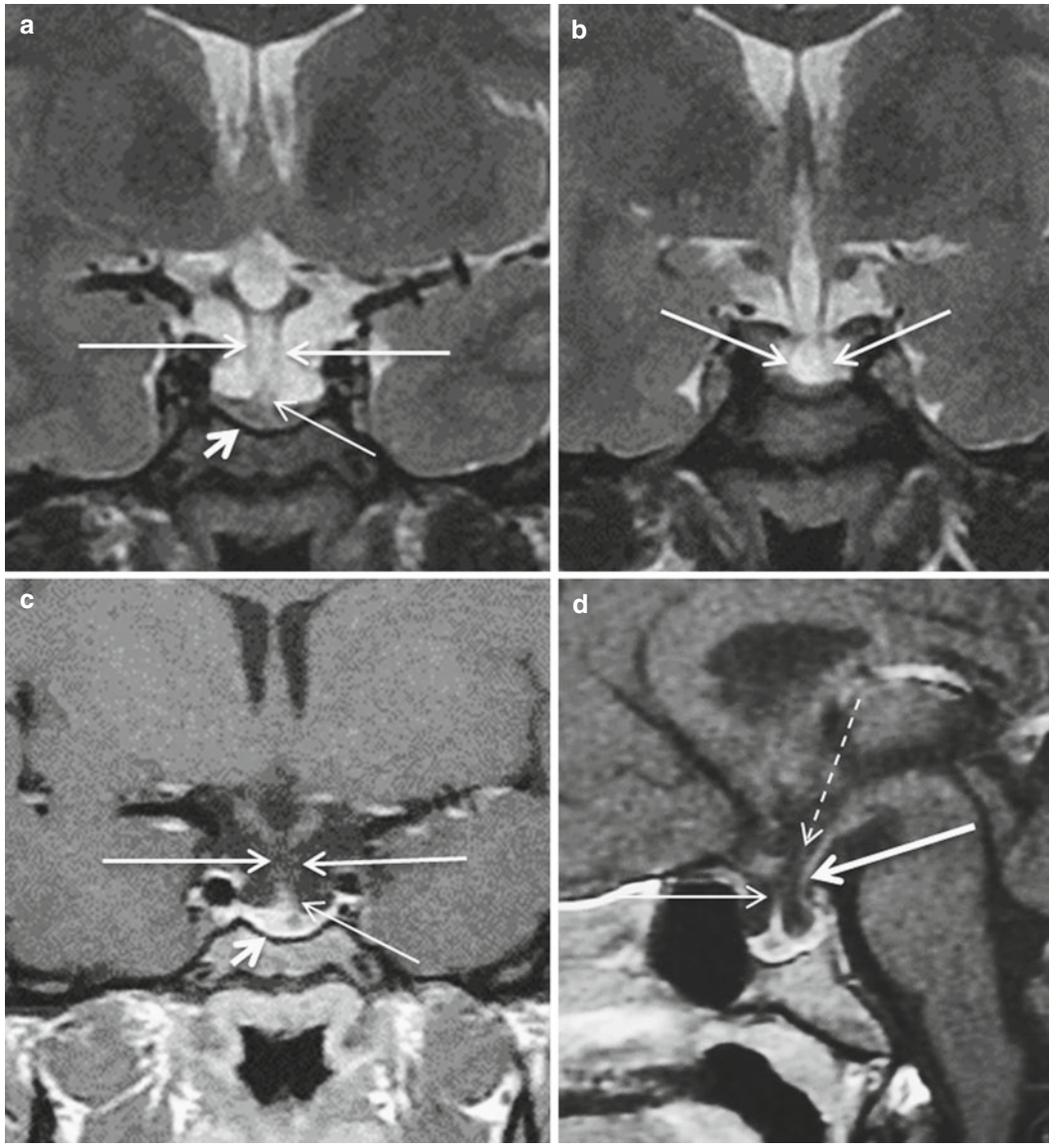

Fig. 57.2 A 20-year-old woman with harmonious dwarfism, spanomenorrhea, and dysmorphism. Pituitary stalk triplication. (**a**, **b**) Coronal T2WIs. Two separate parasagittal pituitary stalks, extending inferiorly to the lateral parts of the pituitary gland (*thick arrows*). Pituitary gland insertion of a median pituitary stalk (*thin arrow*). Single hypoplastic pituitary gland within a large sella (*short arrow*). (**c**) Coronal and sagittal CE T1WIs. Two separate parasagittal pituitary stalks (*thick arrows*). Enhancement of the median pituitary stalk (*thin arrow*) and pituitary gland (*short arrow*). (**d**) Midline sagittal CE T1WI. Defect at the anterior aspect of the floor of the third ventricle (*dotted arrow*) enlarging the infundibular recess without mass effect. Median anterior pituitary stalk (*thin arrow*). Left parasagittal pituitary stalk (*thick arrow*)

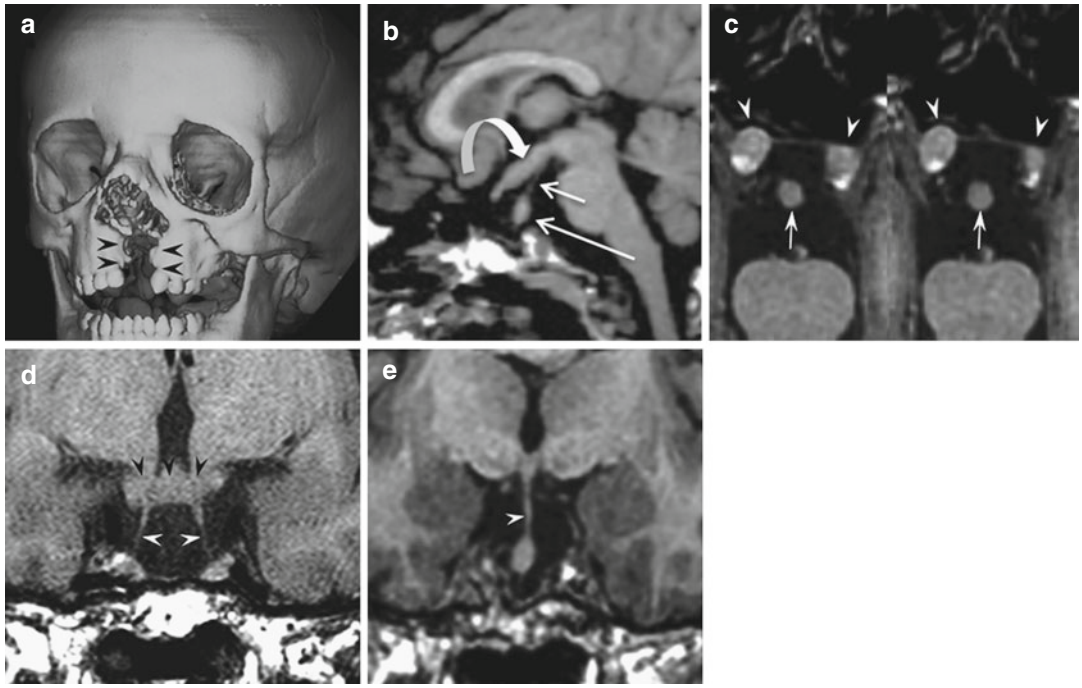

Fig. 57.3 7-y-old first child with midline palate cleft, occult spinal dysraphism, scoliosis. Hypophyseal triplication. (**a**) CT, 3D reconstructed image of the head : midline palate cleft (*black arrowheads*). (**b**) Midline sagittal multiplanar reconstruction T1WI. Thickened third ventricle floor (*curved arrow*). Homogeneous isointense median mass (*long arrow*) whith its independent stalk (*short arrow*). (**c**) Axial multiplanar reconstruction T1WIs: 3 pituitary glands are contemporaneously visible: two parasagittal each with a neurohypophyseal bright spot (*white arrowheads*) and a median one (*arrow*). (**d**) Coronal T1WI. Paired infundibula (*white arrowheads*) extending inferiorly to two pituitary glands fossa. The hypothalamus appears grossly thickened with two infundibula (*black arrowheads*). (**e**) Coronal multiplanar reconstruction T1WI : third medial pituitary stalk (*white arrowhead*). (Courtesy of R.Manara, MD)

Further Reading

De Penna GC, Pimenta MP, Drummond JB et al (2005) Duplication of the hypophysis associated with precocious puberty: presentation of two cases and review of pituitary embryogenesis. Arq Bras Endocrinol Metabologia 49:323–327

Kandpal H, Seith A, Philip J et al (2007) Partial duplication of the hypophysis in adult patients: report of 2 cases. J Comput Assist Tomogr 3:365–367

Manara R, Citton V, Rossetto M et al (2009) Hypophyseal triplication: case report and embryologic considerations. Am J Neuroradiol 30:1328–1329