TITLE: Newborn with Duplication of the Pituitary Gland (DPG)-plus syndrome and congenital hairy polyp.

ABSTRACT:
Ex-39w2d female newborn with prenatally-diagnosed left-sided congenital diaphragmatic hernia with resultant pulmonary hypoplasia (s/p thoracoscopic repair), cleft palate with a nasal/ oral mass, agenesis of the corpus callosum, and duplication of the pituitary gland (DPG)-plus syndrome. Otolaryngology was consulted on DOL 6, where physical exam showed a midline nasal mass visualized through a wide cleft of the secondary palate and a midline nasal dorsal pit. CT and MRI imaging revealed a duplicated pituitary gland, corpus callosum agenesis, and a sizable mass protruding through the cleft palate, exhibiting features most consistent with congenital nasopharyngeal teratoma. Initial plans were to delay resection until the baby was older, however both feeding and respiratory support plateaued, and there was concern the mass was impacting feedings. The infant was taken to the OR for resection using combined trans-oral and trans-nasal endoscopic approach on DOL 28. Intraoperatively, a pedunculated mass arising from the posterior tongue was also resected. Pathology revealed a lingual hamartoma, and the large nasopharyngeal mass was consistent with a congenital hairy polyp. Postoperatively, feedings did not improve, and placement of a gastrostomy tube was ultimately recommended.

A literature search on PubMed did not reveal any case reports of newborns with DPG-plus syndrome and concomitant congenital hairy polyp. In total, there have been approximately 50 cases of DPG-plus syndrome reported worldwide, a small number of which have had associated nasopharyngeal teratomas. A handful of hairy polyps have been reported separately, originating within the nasopharynx, esophagus, and eustachian tubes, among other locations.