

Congenital tracheoesophageal, umbilical, vascular and meningeal fistulas

Abstract

Tracheoesophageal fistula and esophageal atresia are rare congenital abnormalities which represent two components of VACTERL association of anomalies. They are frequently associated with laryngeal cleft. Congenital umbilical fistulas result from failure of obliteration of the fetal vitellointestinal duct or urachus. Vitellointestinal duct abnormalities are rare and they include Meckel's diverticulum, vitelline cyst and vitellointestinal fistula. Urachal anomalies are very rare, and they include diverticula, cysts, sinuses and fistulas. Ultrasonography and fistulography are helpful investigations for the diagnosis of urachal fistulas; the fistula is surgically excised together with the adjoining apex of the urinary bladder. Congenital prepubic sinuses and urethroperineal fistulas are other rare anomalies. Congenital coronary artery fistulas may be also discovered between the coronary arteries and the four cardiac chambers or the great vessels of the heart. Coronary arteriovenous fistulas and absent ductus venosus with abnormal venous circulation could occur as rare congenital anomalies. Other rare congenital arteriovenous fistulas could exist in the thorax, abdomen, pelvis, and cranial and spinal meninges.

Keywords: congenital, fistulas, trachea-esophageal, vitellointestinal, urachal, arteriovenous, meningeal

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Abbreviations: AVF, arteriovenous fistula; CPS, congenital prepubic sinus; CUFs, congenital umbilical fistulas; EA, esophageal atresia; GERD, gastroesophageal reflux disease; HUC, hernia of umbilical cord; IVC, inferior vena cava; OMD, omphalomesenteric duct; TEF, tracheoesophageal fistula; VID, vitellointestinal duct

Introduction

Fistula is a pathological tract connecting two internal cavities or an internal cavity with body surface. In addition to congenital fistulas of the head and neck which we have previously reviewed¹ congenital tracheoesophageal, umbilical, arteriovenous and nervous fistulas are described here.

Tracheoesophageal fistula and VACTERL association

Esophageal atresia (EA) is a rare congenital anomaly (1:2500-4500 live births) which is most frequently associated with tracheoesophageal fistula (TEF). TEF and EA are two components of VACTERL association of congenital anomalies: "V" for vertebral or vascular anomalies, "A" for anal atresia, "C" for cardiac abnormalities, "T" for tracheoesophageal fistula, "E" for esophageal atresia, "R" for renal abnormalities, and "L" for limb anomalies.^{2,3} VACTERL is an association rather than a syndrome as its components are not pathologically caused but embryologically formed due to mesodermal defects.⁴ When at least two⁵ or three^{6,7} components of these congenital anomalies are identified in the same neonate, VACTERL association is diagnosed. Patients with EA most likely suffer from gastroesophageal reflux disease (GERD).⁸

Children with EA/TEF have a greater incidence of laryngeal cleft than other free children.⁹ Neonatal surgical repair of ET/TEF is usually lacks sufficient preoperative information about the anatomy of these structures.¹⁰ Postoperative complications of EA/TEF surgical repair

include recurrence, intestinal sequestration, and repeated thoracic drainage procedures.¹¹

A rare congenital esophageal diverticulum in children is commonly non-symptomizing; it may be complicated with a bronchoesophageal fistula leading to chronic cough due to recurrent pneumonia. Anterolateral thoracotomy is adopted to excise the diverticulum and close the fistula.¹² In an adult woman, a rare congenital bronchoesophageal fistula with intralobar pulmonary sequestration was thoracoscopically excised with favoring results.¹³

Congenital umbilical fistulas

Congenital umbilical anomalies result from persistent patency of the vitellointestinal duct (VID), the allantois (urachus) or the umbilical ring. Congenital umbilical fistulas (CUFs) result from failure of obliteration of either of the VID or the urachus. They are identified postnatally when signs of umbilical pathology: swelling, redness, or discharge are observed.¹⁴ Meckel's diverticulum is the commonest congenital anomaly of the gastrointestinal tract; it occurs in 1-3% of autoptic specimens.¹⁵ In early development, the fetal VID (omphalomesenteric duct: OMD) connects the midgut loop with the yolk sac; with further development it is obliterated and degenerated leaving no remnants. Partial or complete patency of VID results in Meckel's diverticulum, vitelline cyst or vitellointestinal fistula; the latter is associated with persistent fecal umbilical discharge dated since birth.¹⁶⁻¹⁹ A patent OMD is commonly connected to the ileum but rarely to the caecum or vermiform appendix.²⁰ Few cases of rare neonatal congenital umbilical vermiform appendices with appendico-umbilical fistulas are reported. VID fistulas may be also iatrogenic in etiology due to faulty clamping and severing of umbilical cords containing herniated midgut loops.^{21,22} Few cases of rare congenital fistulation of Meckel's diverticulum to the surface of an intact exomphalos sac are reported.^{23,24} A case of a child discharging a roundworm through the umbilicus was described as a strange presentation of a patent VID.²⁵

Another case of a congenital umbilical fistula discharging bile was also discovered in a full-term neonate; it communicated with the intrahepatic biliary system.²⁶ Closure of the congenital umbilical intestinal fistulas in children is recommended as persistence of fistula results in the formation of intraabdominal adhesions.²⁷

The allantois (allanto-enteric diverticulum) is an endodermal diverticulum extending from the ventral part of the cloaca (termed the primitive urogenital sinus) toward the umbilical ring. The cloaca is the dilated caudal end of the hindgut. With further development, the allantois becomes narrowed and termed the urachus of which the proximal part forms the apex of the urinary bladder whereas the distal part becomes obliterated and persists as the median umbilical ligament.²⁸ Congenital urachal anomalies: cysts, fistulas, sinuses, and diverticula are very rare.²⁹ Urachal abnormalities result from either incomplete fetal obliteration of the urachus or postnatal regression leading to recanalization of the urachus after its closure.³⁰ Fistulography and ultrasonography are valuable diagnostic methods and complete urachal excision is the treatment of choice.^{31,32} Congenital patent urachus in adults is rarely reported; in a middle-aged Japanese male a urachal fistula was resected together with partial cystectomy.³³ It is advisable not to excise the umbilicus in such cases, and laparoscopic surgery seems to be of satisfactory outcome.³⁴

A congenital prepubic sinus (CPS) is another rare abnormality that is supposed to be due to persistence of a part of the cloacal membrane.³⁵ A case of midline CPS was described to extend from the prepubic skin to the superior surface of the urinary bladder, then to the umbilicus along the median umbilical ligament.³⁶ In a boy suffering from abnormal dribbling of urine from his perineum, a congenital posterior urethroperineal fistula was extending from the posterior urethra to the perineal skin.³⁷ However, a urethrocutaneous ventral fistula of the penis might be iatrogenic due to traumatic injury during a previous circumcision.³⁸ Anterior perineal sinuses along the median raphe are supposed to be due to congenital midline inclusion disorders³⁹ whereas pilonidal sinus and pyoderma fistulans sinifica are considered as retention skin lesions and not of congenital origin.⁴⁰

Congenital vascular fistulas

Congenital coronary artery fistulas are abnormal communications between the coronary arteries and the four cardiac chambers or the great vessels of the heart. Rare coronary arteriovenous fistulas are often congenital but may be iatrogenic.^{41,42} Microfistulas were detected between the left anterior descending artery and the left ventricular cavity on cardiac catheterization of an old patient.⁴³ Rare origin of the left main coronary artery from the right Valsalva sinus is a congenital anomaly that is commonly associated with coronary fistulas and other cardiac abnormalities; such coronary anomaly is described in the literature as a rare association with Marfan syndrome.⁴⁴ An uncommon combination of congenital coronary anomalies (atresia, stenosis and fistula) in a child was diagnosed and surgically managed.⁴⁵

Ductus venosus is a normal fetal shunt between the left branch of the portal vein and the inferior vena cava (IVC). It is obliterated at birth to form the ligamentum venosum of the liver. Rare absence of the ductus venosus may be seriously associated with abnormal umbilical vein that bypasses the liver (and IVC) and directly terminates into the right atrium.⁴⁶

Congenital arteriovenous fistula (AVF) is extremely rare. An AVF is reported between the left internal thoracic (mammary) artery and the ductus venosus.⁴⁷ A rare neonatal congenital AVF between the inferior

epigastric arteries and the umbilical vein had resulted in a clinical condition simulating congestive heart failure; the affected vessels were surgically ligated and excised.⁴⁸ A rare congenital systemic-pulmonary AVF, manifested by pulmonary hypertension, was detected on using aortic angiography and echocardiography.⁴⁹ An extremely rare case of congenital intrahepatic AVF was prenatally diagnosed.⁵⁰ A fistula between the internal thoracic artery and the pulmonary vessels is a rare congenital anomaly, it could be also iatrogenic in case of coronary artery surgery, trauma or neoplasms.⁵¹ A congenital pulmonary AVF is mentioned to be a risk factor for embolic stroke.⁵² An extremely rare congenital fistula between the right pulmonary artery and the left atrium may cause cardiac failure in utero.⁵³ Congenital aortocaval fistula between the right sinus of Valsalva and the superior vena cava is a very rare anomaly that causes left to right shunt.⁵⁴ In the rare congenital aorto-atrial fistulas, abnormal connections exist between the different parts of aorta and the atria leading to volume overload and heart failure.⁵⁵ AVF between the inferior mesenteric artery and vein is very rare and it may cause ischemic colitis.⁵⁶ An extremely rare case of an ilio-iliac AVF is also reported in the literature with atypical clinical manifestations.⁵⁷

Congenital fistulas in the nervous system

Arteriovenous fistulas are rarely encountered in the nervous system. A congenital cranial pial AVF is extremely rare. A pial AVF was detected originating from the left middle cerebral artery and terminating into the superior sagittal venous sinus.⁵⁸ Also, there is a report of a prenatally diagnosed intracranial pial AVF ending into the great cerebral vein of Galen; it was treated in the early neonatal life.⁵⁹

Spinal cord arteriovenous fistulas may exist in the spinal cord itself or in the spinal meninges. They include the spinal dural AVF as the commonest and acquired lesion and the spinal pial AVF as the less common and congenital lesion. The co-existence of different types of spinal cord arteriovenous shunts is very rare.⁶⁰ Concomitant existence of spinal lipoma and acquired spinal AVF is also very rare. The lipoma releases angiogenic factors that are involved in the development of the spinal AVF.⁶¹

Conclusion

An adequate knowledge of the pathoembryology of the congenital diverticula, cysts, sinuses and fistulas, at different body regions, is essential for proper prenatal and early postnatal diagnosis. Early intervention in such cases is necessary for minimizing postoperative complications.

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Conflict of interest

The author declares no conflict of interest.

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