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Short Communication

Ontogenesis and Imbroglio: The Circinate Pancreas

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Abbreviations

PV: Portal Vein; SMV: Superior Mesenteric Vein; SMA: Superior Mesenteric Artery; SV: Splenic Vein

Initially categorized by Tiedeman in 1818, Annular pancreas (Annularis –alluding to ring shaped in latin) is a sporadic, congenital anomaly of the pancreas. Though periodically inert, annular pancreas may clinically evolve into an extensive infirmity. Classic neonatal intestinal obstruction is expressed besides intricate and composite complications in the adults. Annular pancreas is a significant structural and anatomical configuration of neonatal intestinal obstruction ,which requires early recognition and appropriate therapy. The malformation is considerable, as detected in the precocious phase and prenatal assessment is expanding substantially.

Probability and Analysis: This preponderantly male congenital aberration occurs with a prevalence rate of 1-3 in 20,000 [1,2]. The disorder can present spasmodically and asymptomatically and therefore the presentation may be post-mortem. Nevertheless, an appreciable number of patients are discovered in the prenatal or early life. Infrequently, the disorder remains undisclosed till adulthood. Annular pancreas can be diagnosed at an unspecified period (the age of exposition reveals a bell shaped curve) and may be discerned either accidentally or secondary to inflammatory insults such as acute pancreatitis. Symptoms in adulthood are generally postponed till the 3rd or 6th decade [3]. Annular pancreas has a powerful alliance with coexisting congenital aberrancies in up to 71% cases. The most constant coalition is with the Down's syndrome, although miscellaneous forms of Cardiac and Gastrointestinal congenital malfeasance are enumerated incorporating Hirschsprung's disease, Imperforate anus, Trachea-oesophageal fistula and Oesophageal atresia along with pathologies such as Pancreatic cancer, Pancreatitis, Pancreas divisum and Intraductal papillary mucinous neoplasia (IPMN).

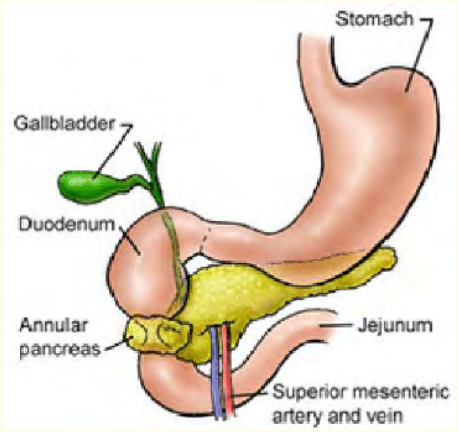

Figure 1: *Diagrammatic representation of annular pancreas.*

Analysis and Examination: Annular pancreas is an embryological deformity of the foregut. Evolution of the pancreas commences in the fifth week of gestation. One dorsal and one ventral bud expands from the primitive foregut. The ventral bud rotates along with the gut by week seven to coalesce with the dorsal buds after trailing in the wake of the duodenum. The dorsal bud configures the body and the tail of the pancreas and the ventral bud constructs the inferior part of the pancreatic head and the uncinate process. Amalgamation of the buds designs the main pancreatic duct. The hypothetical premises indicated for the evolution of annular pancreas are i) The tip of the ventral bud demonstrates an atypical coalition with the duodenum with erroneous rotation, thereby terminating in a band of fibrous or pancreatic parenchymal tissue wrapped around the second part of the duodenum. ii) Alternatively, it is assumed that hypertrophy of the dorsal and the ventral buds ensue in an intact band of pancreatic tissue surrounding the duodenum and an imperforate ring of pancreatic tissue is structured in almost 25% cases [4]. The ventral buds are hindered from displacement with the duodenal rotation, which bring about the encirclement of the descending part of the duodenum. The circumscription is categorized into a) complete – the variant which leads to complete duodenal obstruction. b) incomplete- where the anterior duodenum is exposed and which elucidates symptoms substructured on inflammation or alternative pathologies.

Clinical Demonstration

History: Annular pancreas can be detected in the prenatal phase due to a) complications potentiating as emergencies, b)coincidentally after imaging procedures, c) during surgical intervention or d) at autopsy. The band of pancreatic tissue constricting the duodenum induces the clinical symptoms in the first few hours after birth. Neonatal intestinal obstruction may principally present in a vague manner constituting of poor feeding, vomiting and irritability. Obstruction proximal to the ampulla of vater induces a non-bilious kind of vomiting which can be misinterpreted as mild, non-life threatening conditions. The age of the patient predominantly defines the clinical scenarios though the disease customarily appears in infancy or early childhood. Generally the affected children display gastro-intestinal symptoms such as poor feeding , vomiting and abdominal distension. Delayed representation demonstrating malnutrition, failure to thrive, bile stained vomiting and abdominal cramps is exhibited in Africans. Adults usually depict upper abdominal pain or acute pancreatitis. Annular pancreas is an infrequent source or a paediatric neonatal duodenal obstruction. However a high index of suspicion is obligated, in order to establish the diagnosis and institute appropriate and early therapy. Adult patients, generally discovered between 20 - 50 years, elucidate abdominal pain, nausea, vomiting, weight loss and obstructive jaundice. Annular pancreas is detected in males more than in females. In 25% individuals , a circumferential pancreatic enclosure is delineated while 75% display a partial ring. With the onset of obstructive jaundice, a suspicion of a carcinoma of the ampulla of vater arises [5]. Annular pancreas manifesting in the adults are generally affiliated with complications of peptic ulcer, pancreatitis, duodenal and biliary obstruction [6,7]. Congenital anomalies such as Down's syndrome, Esotracheal fistula, Anal imperforation, Hirschsprung' s disease and Smith-Lemli-Opitz Syndrome may also be discovered concurrently with annular pancreas. 40% of the annular pancreas are usually established on laparotomy. Duodenojejunostomy and gastrojejunostomy are preferred in adults because of restricted duodenal flexibility. Laparoscopic gastrojejunostomy is the recommended procedure in the obese and those with gastrointestinal malignancies. Resection of the annular pancreas from the duodenum may demonstrate duodenal leakage, pancreatitis, pancreatic fistula etc.

Physical Assessment: Clinical examination and conclusion primarily depends upon the age of the exposition and the degree of the systemic involvement. When presenting as an intestinal obstruction, the abdominal distension (prior to decompression), fluctuating bowel sounds and a palpable peristalsis is visualized with the initiation of intestinal obstruction. Delayed representation elucidates signs of hypovolemic shock constituting of pallor, poor capillary refill and drowsiness or lethargy.

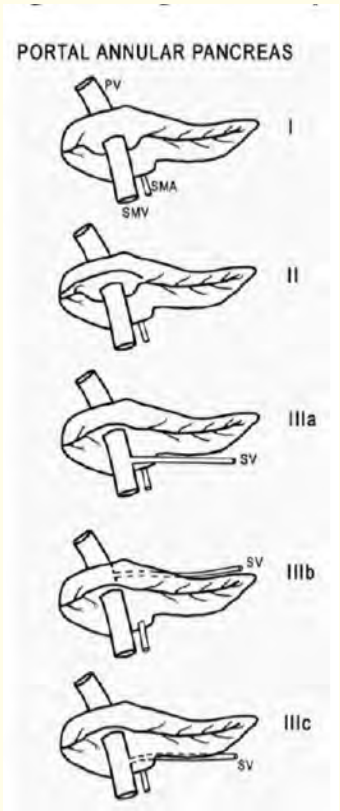

Figure 2: *Classification of AP as per vascular sequence.*

Inquisition: Prenatal diagnosis: Confirmation of the existence of annular pancreas in foetal life (2nd trimester) is now feasible. Prenatal and a vague assessment of duodenal obstruction is possible with the coexistent dilatation of the stomach and the duodenum – the classic radiographic double bubble sign. However this sign is not an unequivocal indication for diagnosing annular pancreas nor does it categorically enunciate the logic for the duodenal obstruction. Annular pancreas generally qualify as only 1% in the case incidence of neonatal duodenal obstruction [2]. Thus definitive pre-natal markers for the discernment of annular pancreas needs corollaries. Hypoechogenic bands observed around the duodenum characterize the existence of annular pancreas [4]. The subsistence of annular pancreas in foetal life can also be detailed by the hypoechogenic bands.

Manner of Imaging: Definitive and architectural erudition of the pancreas is not available on plain radiography of the abdomen on account of the absence of tissue contrast. Imaging modalities incorporating the cross sectional techniques are beneficial for the accurate anatomical exposition of the pancreas. Notwithstanding, total duodenal obstruction classically delineates a double bubble on abdominal radiograph. Conventionally it symbolizes dilatation of the stomach (exhibited in the left upper quadrant) and proximal duodenum (expressed in the right upper quadrant). Abdominal radiography depicting a double bubble is generally enough to diagnose a complete duodenal obstruction. Notwithstanding an upper intestinal contrast radiograph is conducive for distinguishing a partial duodenal obstruction.

Ultrasonography: Appraisal with an ultrasound permits real time estimation of the paediatric pancreas, in the absence of ionizing radiation. and is usually without the need of sedation. The procedure cogitates a protective, inexpensive and an accessible modality for imaging the paediatric pancreas. Children display an optimal anatomic window provided by the liver in addition to the diminished thickness of the abdominal surface, which inspires the acquisition of an enhanced quality of the pancreatic images [8].

Computed Tomography: The slender band of pancreatic tissue around the duodenum in combination with minimal intra-abdominal fat makes the comprehension of the paediatric annular pancreas with the computerized tomography (CT) a challenging methodology. Nevertheless, the disorder is consistently discovered with the established intravenous (IV) contrast enhanced pancreatic study [8,9]. CT can also emphasise the extraneous compression of the duodenum in contradiction to the consolidation of the duodenal wall. Aberrations discerned by the ultrasound can be extensively assessed by the CT.

Magnetic Resonance Imaging (MRI): MRI is an addendum to the CT imaging for the interpretation of anatomy of the pancreas Compounded with Magnetic Resonance Pancreatography (MRCP), it permits imaging of the specific aspects of the biliary system [8]. Conventionally, the precise nature of biliary duodenal obstruction is not appreciated until adequate surgical exploration is executed.

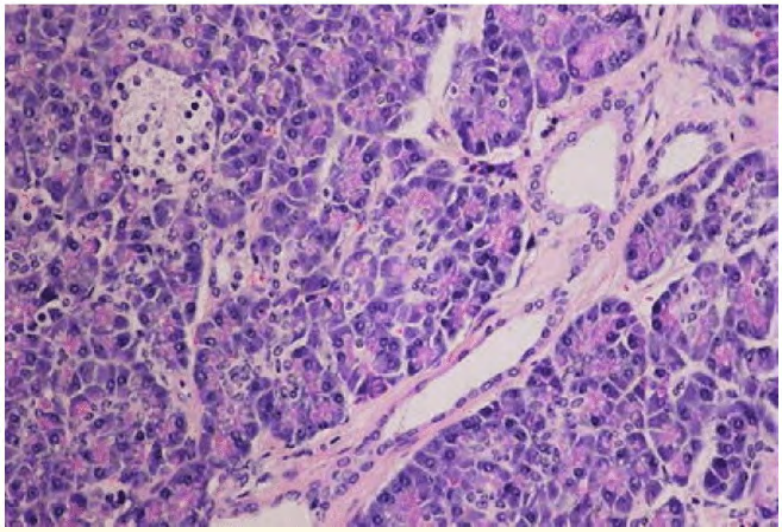

Figure 3: *Exocrine Annular Pancreas.*

Management: The asymptomatic cases of annular pancreas entail minimal therapeutic intervention. Specialized management and treatment is subject to the clinical symptoms engendered by the congenital aberration or the type of complications generated. Eventually duodenal obstruction demands immediate surgical intervention even though the exact basis of the obstruction is obscure. However, the therapeutic regimen should incorporate abdominal decompression with a nasogastric tube and resuscitative measures fortified by fluids and electrolytes. The patients can be relocated to high dependency, specialized care units where appurtenant monitoring of the surgical intervention deemed necessary can be initiated. Therapeutic deferment may culminate in a serious systemic chaos thus it is imperative that appropriate resuscitation with fluid and electrolyte compensation is instituted prior to surgery. Duodenal obstruction secondary to detected annular pancreas is managed by surgical bypass constituted of duodenoduodenostomy or duodenojejunostomy. If the genesis of the obstruction is not apparent on pre-operative imaging, an exploratory laparotomy is indicated. With gross dilatation of the proximal segment, the tapering of the bowel is optional. Transanastomotic nasogastric tubes are inevitable where parenteral nutrition is not accessible, or the enteral feeding is interrupted on account of the dilated proximal segment.

Consequences and Predictions: Expeditious recognition, resuscitation, and a clearly defined management protocol forms the basis of efficacious treatment outcomes in neonatal intestinal obstruction. Extraordinary technical skill is imperative to bypass the obstruction detailed by the aberrant pancreatic tissue. Appropriate peri- operative care is essential to assure a favourable surgical outcome. Surgical intervention of the annular pancreas has excellent results [10] when accompanied with pertinent peri- operative and postoperative care. Befittingly trained anaesthetic staff, ventilator support and dynamic parenteral nutrition [11] is an essential component of the care. However the surgical eventualities may be compromised in the event of severe concomitant congenital anomalies.

Prohibitions and Corollaries: Prenatal diagnosis along with the analysis of pertinent markers for the appraisal of annular pancreas have ameliorated -thus the discovery of the congenital anomaly is excellent, though it cannot be anticipated. Initial assessment of complications especially intestinal obstruction should confirm that suitable therapy is to be initiated at the earliest. Surgical intervention for duodenal obstruction ensures commendable prognosis, although the comprehensive outcome is also dependent upon the coexistent, severe congenital aberrations [12-18].

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14. Image 1 Courtesy: Medbullets.
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