Dorsal pancreatic agenesis is an extremely rare anomaly which results from an embryological failure of the dorsal pancreatic bud to form the body and tail of the pancreas. During the last 100 years in medical literature, there are only 54 reports with clinical descriptions of agenesis of the dorsal pancreas in humans. Agenesis of the dorsal pancreas is mostly asymptomatic but abdominal pain, pancreatitis and diabetes mellitus may clinically be apparent. We present a case of dorsal pancreatic agenesis detected incidentally on imaging.

CASE REPORT
A 30-year female patient presented to our hospital after right hemicolectomy for carcinoma (CA) colon. Patient was having history of vague right sided abdominal pain for the last one year, which was aggravated for the last few days. She had consulted some private hospitals where USG abdomen was performed. USG revealed a gut related mass involving the ileocecal region. She underwent laparotomy. Peroperatively, there was an ileocolic intussusception associated with malignant looking cecal mass. Right hemicolectomy with ileocolic anastomosis was performed. Histopathology revealed poorly differentiated adenocarcinoma arising from cecum with signet ring cell features. All margins were free of tumor, and 13 lymph nodes removed were all negative for metastatic carcinoma. Patient presented to our hospital for further treatment of CA colon. Postoperative contrast enhanced CT scan abdomen and pelvis was performed in our Department. CT revealed no evidence of residual disease. Anastomotic site was unremarkable. Incidental note was made of complete absence of neck, body and tail of pancreas. Head of pancreas was of normal size and parenchymal attenuation having truncated appearance (Figures 1a and 1b). Distal pancreatic bed was filled by intestine (dependent intestine signs), which was abutting the splenic vein (Figure 2). Other intra-abdominal structures, including the liver, spleen, gallbladder, adrenals, kidneys and inferior vena cava appeared unremarkable. Biochemical evaluation of patient revealed mild elevation of fasting plasma glucose (130 mg/dl). Patient was recently diagnosed of having diabetes mellitus during preoperative work-up (with fasting plasma glucose of 180 mg/dl and HbAlc of 14.4% at that time) and was on insulin. Serum pancreatic amylase and lipase levels were normal. Liver and renal function tests were also within normal limits. Based on the clinical and CT findings, diagnosis of isolated agenesis of the dorsal pancreas associated with diabetes mellitus was made. With reference to CA colon, after complete staging work-up, patient was staged as T3N0M0. Keeping in view the poorly differentiated histopathology of patient's malignancy, she received 5 cycles of chemotherapy (5FU and leukoverin). Patient tolerated the chemo-therapy well and was kept on follow-up. She was on insulin therapy for management of her diabetes with fair control.

DISCUSSION
Pancreas develops from ventral and dorsal endodermal buds originating at the junction of the foregut and mid gut. The ventral pancreatic bud and biliary system arise from the hepatic diverticulum, and the dorsal pancreatic bud arises from the dorsal mesogastrium. After clockwise rotation of the ventral bud around the caudal part of the foregut, there is fusion of the dorsal pancreas (located anteriorly) and ventral pancreas (located posteriorly). Finally, the ventral and dorsal pancreatic ducts fuse, and the pancreas is predominantly drained through the ventral duct, which joins the common bile duct (CBD) at the level of the major papilla. The dorsal duct empties at the level of the minor papilla. Abnormal embryogenesis can lead to developmental failure of the dorsal pancreas, resulting in complete

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Received: December 11, 2015; Accepted: June 22, 2016.
agenesis of the dorsal pancreas. This condition is exceedingly rare. Agenesis of ventral pancreas and complete agenesis of the pancreas are incompatible with life. The exact mechanism and etiology of agenesis of dorsal pancreas is not known. Severe hypoplasia of the pancreas can be associated with mutations involving the HNF1β gene.

Most of the patients with agenesis of dorsal pancreas are asymptomatic, but 92.9% of the symptomatic cases present with epigastric pain, which may be non-specific or secondary to pancreatitis. About half of affected individuals develop diabetes mellitus. Sometimes, the patient may present with steatorrhea or other signs of exocrine insufficiency. Other abnormalities such as heterotaxy, polysplenia syndrome, ectopic spleen, bowel malrotation, coarctation of the aorta, tetralogy of fallot, atrioventricular valvular abnormalities, and total anomalous pulmonary venous connection have also been reported to be associated with agenesis of dorsal pancreas.

In this patient, agenesis of dorsal pancreas was associated with diabetes mellitus, however, no other co-existing anomaly was detected.

Conditions that have clinical pictures similar to that of agenesis of the dorsal pancreas include pseudoagenesis (atrophy of the corpus and the tail of the pancreas secondary to chronic pancreatitis), carcinoma of the head of pancreas (proximal atrophy of the gland), pancreas divisum (absence of fusion or incomplete fusion of the ventral and dorsal pancreatic ducts); and distal pancreatic lipomatosis. These conditions can be differentiated from agenesis of dorsal pancreas by a careful medical history and appropriate imaging studies including ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI, including MRCP) or endoscopic retrograde cholangio-pancreatography (ERCP) and endoscopic ultrasound (EUS).

In the differential diagnosis of pseudo-agenesis, histories of previous abdominal pain, pancreatitis, CT scanning and the serum amylase level may be helpful. Multidetector CT (MDCT) scan is also helpful in differentiating distal pancreas agenesis from distal pancreas lipomatosis. In the absence of distal pancreas, distal pancreatic bed can be filled by stomach or intestine (dependent stomach or dependent intestine signs), which abut splenic vein as in this patient. Same findings can be seen in patients with distal pancreatectomy, however, splenic vein is absent in these patients. In case of distal lipomatosis, abundant fat tissue is observed anterior to splenic vein. Dependent stomach and/or dependent intestine signs on MDCT imaging can allow differentiation of distal pancreas agenesis from distal lipomatosis. Park et al. reported a case having distal fat replacement with absent ductal and acinar cells. Therefore, ERCP or MRCP is not necessary for revealing the major and the accessory duct systems; and dependent stomach and/or dependent intestine signs on MDCT imaging can be diagnostic obviating further diagnostic studies. Treatment is not required in asymptomatic patients.

REFERENCES