Test your knowledge by answering the questions provided along with the two sets of images and case scenarios. The answers and a short description are given on the next page.

Case 1:

Case scenario: A gentle man in his late 50s presented to the OPD. He had been suffering from prolonged general ill health and chronic cough. Currently he is worried by development of gradually progressive swelling over face and neck.

1) What is the clinical diagnosis?
2) What would be the 1st line investigations?
3) What would be the recommended line of treatment?

Case 2:

Case Scenario

- A 30 year old lady with sudden onset, severe intermittent colicky pain abdomen associated with multiple episodes of non projectile, non bilious vomiting for one week. Clinical examination revealed an ill defined palpable mass in the right iliac fossa. Blood investigations were within normal limits. Ultrasound and CT abdomen of the lady are shown above.
- What is your diagnosis after having seen the images?
- What are the significant findings in the images that point to a diagnosis?
- What are the investigations you would order in this patient (adult) and in a child who presents with similar clinical features?
Case 1  Dr. Benjamin Barsouma Mathew & Dr. Tony Bishwas, Harriet Benson Memorial Hospital, Lalitpur, UP

This elderly gentleman has presented with generalised fatigue, chronic cough and recentonset progressive swelling of face and neck. There is oedema of the face and arms associated with plethora and swollen collateral veins on the front and back of the chest wall and arms. An obstruction to the Venous Drainage from upper part of body leading to extravasation of fluid and oedema and opening up of collaterals leading to engorged veins in chest wall explains the clinical picture. The clinical Diagnosis will be that of a Superior Vena Cava syndrome.

Superior vena cava (SVC) syndrome is the constellation of symptoms and signs that results from effective obstruction of blood flow at the level of the SVC itself, the great veins as they empty into the SVC, or the superior cavo-atrial junction. SVC syndrome was first described by William Hunter in 1757 in a patient with a large syphilitic aortic aneurysm compressing the SVC. Because the venous drainage from the upper extremities, upper thorax and head is obstructed, SVC syndrome presents with symptoms related to engorgement of these areas.

The various manifestations of SVC syndrome is summarised in Table 1, the most common ones being facial and neck swelling, dyspnoea and cough.

The causes of SVC syndrome are summarised in Table 2. The most common cause today is a Bronchogenic Carcinoma compressing the SVC. Such a mass is usually visible on a chest X-Ray making it the first investigation of choice. The X Ray of this patient(Fig 1) showed a right upper zone, well defined round homogenous opacity, most probably bronchogenic in origin. A Computed Tomography may be indicated to define the level and extent of venous blockage, map the collaterals and identify and stage the underlying
cause. Conventional superior vena cavography is the gold standard for identification of SVC obstruction and the extent of associated thrombus formation. Investigations for diagnosing and staging the underlying cause are done in parallel.

The goals of management for SVC syndrome associated with malignancy are to alleviate symptoms and treat the underlying disease. Contrary to the earlier belief, not all cases of SVC syndrome require emergency management.

The general principles of management of SVC syndrome are:
- Head elevation to decrease venous stasis
- Preventing injections to arm to avoid thrombosis in presence of pre-existing stasis
- Removal of indwelling catheters, which might have precipitated a thrombus
- Anticoagulation if thrombus is the cause
- Glucocorticoids are found to effective in steroid responsive tumours (e.g., Lymphomas) and RT induced SVC syndrome
- Diuretics to alleviate symptoms are often used, although with questionable benefit
- Chemotherapy, Radiotherapy or Surgery for the primary disease according to the tissue diagnosis and stage (in case of external compression)
- Endovascular stenting, often as an emergency procedure in acute cases or as a palliative procedure.
- A surgical SVC bypass, in specific circumstances, where a stenting is not possible

REFERENCES
2. The Superior Vena Cava Syndrome: Clinical Characteristics and Evolving Etiology, Rice et al; Medicine Jan 2006

CASE 2

Drs. Madhavi. K / Geethu Punnen / Shyamkumar N. K.- Dept. of Radiodiagnosis, CMC Vellore

DIAGNOSIS: Jejuno-jejunal intussusception with lipoma as lead point

INTUSSUSCEPTION
- Intussusception is the most common cause of intestinal obstruction in infants and toddlers. It is defined as telescoping of a segment of bowel (intussusceptum) into a distal segment of the bowel (intussuscipiens). There is usually a ‘lead point’- a malignant or benign mass that pulls one bowel segment into another.
- The most common type is ileocolic, followed by ileoileocolic, ileoileac and colocolic. Jejuno-jejunal intussusception (as in this case) is uncommon.
- In children, there is no defined lead point, commonly associated with lymphoid hypertrophy following a viral illness. Intussusception is rare in adults; 90% of cases will have a demonstrable cause (neoplasms account for 60% of cases, followed by inflammation, trauma, adhesions).

CLINICAL PRESENTATION
- Children present with a triad of colicky abdominal pain, vomiting and red currant jelly stools.
- Adults usually have an indolent course, with recurrent abdominal pain, vomiting and rarely palpable mass.
- Ultrasound is the investigation of choice in children. In adults, ultrasound is the base line investigation, followed by cross sectional examination (CT) to assess the cause for the intussusception.

TREATMENT
Patients will have to be kept nil per oral (NPO), IV fluids have to be initiated. Children will require Barium or hydrostatic reduction, air enema or surgery. Adults will require surgical reduction (in malignancy) or resection without reduction (benign lesion). This lady underwent laparotomy, reduction of intussusception and excision of sub mucosal lipoma.
CME IN IMAGES

IMAGING FINDINGS

- Longitudinal ultrasound image shows thickened, edematous hypoechoic outer bowel - intussuscipliens with echogenic central collapsed, telescoping inner segment – intussusceptum and associated mesenteric fat
- Transverse ultrasound image shows an outer edematous, hypoechoic bowel loop with central collapsed telescoping segment of bowel with associated mesentery giving a target or doughnut sign
- There is colour uptake within suggesting preserved vascularity

![Fig. 1: Longitudinal ultrasound image. Fig.2: Transverse ultrasound image.](image)

1= outline of hypoechoic outer bowel, 2= collapsed inner segment, 3= mesenteric fat

(Compare these images with the earlier images)

![Figure 3: Longitudinal (Fig.3: upper image) and serial cross sectional (Fig.3: lower image) diagrams illustrate intussusception with a lead point.](image)

The thick arrows indicate lead mass. The intussusceptum appears thick with loss of gut signature due to edema.

(Image source: Radiographics 2006;Vol 26: 733-744)

The CT coronal image (Fig.4) shows telescoping of the proximal jejunal loops and along with mesenteric vessels into distal jejunal loop.

CT transverse section (Fig.5) shows well defined fat attenuation lesion - lipoma (3) as lead point

(Compare these images with the earlier images)

![Fig.4: CT coronal section, Fig.5: CT transverse section. Arrows 1= outline of outer bowel loop, 2= collapsed inner bowel, 3- Lipoma as the lead point (Compare with earlier images)](image)