Abstract:
Aim To report a successfully operated case of Carcinoma Oesophagus with Situs inversus totalis. Case Report 54 year old postmenopausal woman was evaluated in our department for her complaint of progressive dysphagia. Endoscopy showed growth lower third oesophagus with biopsy showing it to be squamous cell carcinoma. Further evaluation showed no significant perioesophageal disease or any major vascular involvement or distant metastasis. Her radiological investigations (chest x-ray and CT) showed features of Situs inversus totalis. We decided to proceed with Transhiatal oesophagectomy. Unique Anatomical Findings Peroperatively, all her abdominal viscera were in a mirror-image position. We successfully performed the surgery as planned taking necessary precautions while mobilizing the oesophagus in the superior and middle mediastinum considering the altered anatomy of aortic arch and descending aorta. Postoperative period was uneventful.

Keyword: situs inversus, carcinoma esophagus, transhiatal esophagectomy, right sided aortic arch

INTRODUCTION:
Situs inversus totalis which is a complete mirror-image reversal of all the thoracic and abdominal viscera is a very rare entity which occurs in 1 in 5000 to 1 in 20000 livebirths as against Situs solitus which is normal positioning of all the internal viscera. Carcinoma Oesophagus is one of the common cancers encountered worldwide, its incidence is high in India. The incidence of carcinoma oesophagus is altered neither way in situs inversus totalis. A surgeon, in his lifetime, could possibly see one or two cases of such rare combinations. The uniqueness lies in the management of treating such patients surgically, considering the altered positions of various thoracic and abdominal viscera while performing.
operations like transhiatal oesophagectomy. Unfamiliarity in orientation may lead to unwarranted surprises. In this article we present a case of situs inversus totalis with carcinoma oesophagus for which we were able to successfully perform transhiatal Oesophagectomy anticipating the associated unique anatomical variations that could occur.

CASE REPORT:
A 54 yr old post menopausal female patient presented with dysphagia for the past 3 months. It was progressive in nature, more for solids than liquids. There was no family history of malignancy, no associated co-morbid illness. Patient was a non smoker and non alcoholic. Clinical examination revealed position of apex beat of heart on her right 5th intercostals space. Her laboratory studies did not reveal any abnormality. X-ray chest and ECG suggested features of Dextrocardia (Figures 1,2)
Upper GI endoscopy revealed a circumferential lesion at about 37 to 40 cm from the incisor teeth and the scope could not be passed beyond. Biopsy of the lesion revealed squamous cell carcinoma. Barium swallow (figure 4) and CT scan of the lesion (figure 3) showed a thickening at the lower 1/3rd of the oesophagus extending to the OG junction. There was no vascular encasement or perioesophageal disease. Apart from that all the organs were found to be malpositioned as mirror images. Her performance status was normal. Investigations revealed no evidence of distant metastases. After thorough evaluation of her cardiac status and lung function she was planned for a transhiatal oesophagectomy. An upper mid-line laparotomy was done. There were no liver metastases, ascites, peritoneal metastases, or Paraortic nodes. A 3X4 cm tumor was present in the OG junction. All the internal organs were found to be positioned as mirror images; Liver on the left side, spleen on the right side, greater curvature of the stomach was found on the right side. Multiple adhesions in the omentum that was present were released carefully. Stomach was mobilized. Left gastroepiploic vessels near the spleen identified ligated and divided as the conduit is based on the left gastro epiploic and left gastric vessels. (Conventional: right gastroepiploic and right gastric vessels). Peritoneum over the OG junction was normal. Both crura of the diaphragm were divided. Oesophagus was mobilized around the lower mediastinum after dividing the anterior and posterior vagi. There was a right-sided aortic arch which posed no problem in our mobilization. By a hockey-stick incision made on the right side of the neck, oesophagus was identified and mobilized all around the upper mediastinum till the upper limit of cervical incision. Cervical oesophagus was divided with a good cuff of mucosa and sub mucosa. Greater curvature of stomach was tubed with 75 mm stapler and the specimen was removed via abdomen.(Figures 5,6) Stomach tube was pulled in to the neck and an Oesophago-gastric anastomosis was done in two layers in the neck on its right side (conventional: left side). Post-op period was uneventful and it was managed in a routine way.
Her post-op histopathological report revealed, Infiltrating keratinizing Squamous cell carcinoma extending up to the external muscle coat; resected margins were free; 1/7 nodes showed tumour deposits; therefore she was planned for adjuvant chemotherapy During her first cycle of platinum based chemotherapy, patient developed multiple cerebral infarcts which was managed conservatively by a neurologist and recovered completely.
after which she completed her full course of adjuvant chemotherapy. No adjuvant RT was given. Patient was on regular follow-up till one year post surgery when she developed mediastinal and paraaortic nodal recurrence for which she was referred to pain and palliative care clinic for further management.

DISCUSSION:

Incidence of situs inversus totalis reported in literature varies from 1 to 8000 to 1 in 20,000 live births (10). There is a slight male preponderance Situs inversus totalis is classified into Situs inversus with dextrocardia or levocardia according to the position of apex beat although the heart chambers are inverted in both the groups. Incomplete reversal of viscera is known as Situs Ambiguous. The risk of cardiac malformation is much higher in ambiguous (80%) than in inversus (3-6%) or solitus (0.6%).

In situs inversus, both thoracic and abdominal organs are inverted but their normal anatomical relations including the antero-posterior relationships are maintained. Variations found include dextrocardia, left sided liver, right sided spleen, a left lung with 3 lobes and a right lung with 2 lobes. Other anomalies are asplenia. Polysplenia, various cardiac malformations, vascular malformations and renal malformations (3). When Situs inversus totalis is associated with primary ciliary dysfunction it is known as Kartagener’s Syndrome and these patients develop bronchiectasis and infertility.

Most of the patients with situs inversus remain symptomless and have normal life expectancy unless when associated cardiac malformations. Diagnosis is often made accidentally on routine physical and radiological examination. Typical findings in radiological examination include right sided heart and left sided liver. The axis in the Electrocardiogram will be deviated towards the right.

Incidence of various diseases in these patients is similar to general population apart from cardiac anomalies. The challenge to clinicians especially surgeons arise from the unfamiliar anatomical relationship of the internal organs. With situs ambiguous, associated anomalies are more resulting in more intraoperative difficulty. A thorough preoperative imaging and cardiac evaluation is mandatory to avoid unnecessary surprises during surgery (2, 6). Situs inversus totalis may not be necessarily associated with inversion in autonomic innervation and this may result in strange referred pain pattern and other symptoms. **Situs inversus totalis and malignancy**

Literature review does not suggest an increased incidence of any type of malignancy in situs inversus. Most are single case reports of various malignancies. Gastric cancer, hepatocellular carcinoma, pancreatic carcinoma and various lymphomas are the most commonly reported malignancies. There are about 10 cases of esophageal cancers reported in world literature but none from India (8,9).

The major problem faced during surgery in most of these cases is in dealing with right sided thoracic arch in the superior mediastinum(2). There are few reports in which surgeons have utilized a left thoracotomy successfully (2). Some surgeons have successfully resected the tumor by transhiatal approach(3). There is one report in which a combined hand assisted laparoscopy and thoracoscopy were used to resect the tumor(3). A precise preoperative evaluation preferably with MRI to understand the anatomical variations in this group of patients is mandatory to
avoid unwarranted peroperative complications (1).

**Adjuvant therapy in squamous cell carcinoma Esophagus:**
In patients with squamous cell cancers of the esophagus who have undergone an R0 resection can be observed irrespective of their nodal status or enrolled in clinical trials if node positive. Patients with microscopic or macroscopic residual disease after esophagectomy and with no distant metastases should be treated with fluoropyrimidine based post operative chemoradiation.(11,12)

**CONCLUSION:**
Most of the patients with situs inversus remain symptomless and have normal life expectancy unless when associated cardiac malformations. There is no evidence of increased incidence of any type of malignancy in situs inversus. But when encountered, the surgeon must be aware of the unique anatomical variations that could occur while performing major surgeries in these patients to avoid unnecessary catastrophes.

**REFERENCES:**

Figure 1 Chest X Ray

Figure 2 ECG Figure 3A CT Scan Figure 3B CT Scan Figure 4: Barium swallow Figure 5A Intraoperative photo
Figure 5B Intraoperative photo
Figure 5C Intraoperative photo
Figure 6A Surgical Specimen

Figure 6B Surgical Specimen

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