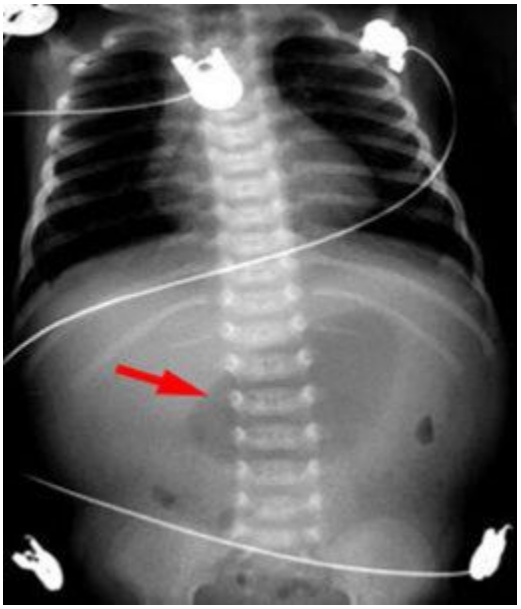
Case of the Week

Case Briefing



A 4 week old full term male child presents to the paediatric surgery OPD with a history of projectile non-bilious vomiting for the past 3 days.The episodes of vomiting follow each feed and the baby is dehydrated and eager to eat again after he vomits.On examination visible peristaltic waves can be seen from left to right in the epigastric region along with a palpable mass in the the right upper quadrant of the abdomen.Plain Xray of the abdomen gives the following picture as shown in the image.

1.What is your provisional diagnosis ?

Answer- The most likely diagnosis here is "Congenital Hypertrophic Pyloric Stenosis"

In the above condition there is gastric outlet obstruction due to thickening of the pylorus which leads to projectile non-bilious vomiting everytime the child feeds.Due to repeated bouts of vomiting the child is dehydrated.Due to the obstruction at the pylorus there is distension of the stomach and visible peristaltic waves of the stomach can be seen moving from left to right.The pyloric thickening can be palpated in the epigastric or right hypochondriac region and is felt as an olive shaped mass.

The given Xray shows the classical "Single Bubble Sign" which is nothing but the distended stomach on X Ray.

USG W/A is diagnostic,pyloric canal length more than 16 mm and pyloric wall thickness of more than 4 mm is significant.

2.Can this condition be suspected in-utero if the pregnant mother has polyhydramnios?

Answer- Even though the name of the disease has the word "Congenital" in it,the disease usually develops over 4 to 6 weeks after birth.Hence the obstruction does not develop in utero.However in certain rare cases CHPS has been documented in in-utero as well wherein the pregnant mother might present with polyhydramios.

In contrast congenital conditions such as annular pancreas and duodenal atresia the obstruction develops in-utero. Hence the baby is unable to swallow the amniotic fluid,which leads to its accumulation.This accumulation of amniotic fluid can be seen clinically as polyhdramnios in the pregnant mother.

3.What metabolic abnormality would be present in this patient? Explain its pathophysiology.
Answer-The metabolic abnormality present in this patient is "Hypochloremic Hypokalemic Metabolic Alkalosis with paradoxical aciduria"

Now due to repeated bouts of vomitting there is loss of hydrogen ions and chloride ions as HCL is present in the stoamch.xNow this loss of hydrogen ions leads to alkalosis and loss of chloride ions leads to hypochloremia.

Now to combat the alkalosis the renal compensatory mechanism is switched on which results in loss of bicarbonates in the urine.Now the bicarbonate ions cannot be excreted alone so it grabs the sodium ions along with it resulting in hyponatremia. This hyponatremia switches on the RAAS system resulting in release of aldosterone which absorbs sodium ions in exchange for potassium ions at the collecting duct initially which again results in hypokalemia. Once hypokalemia develops sodium ions are exchanged with hydrogen ions to prevent further hypokalemia.This excretion of hydrogen ions in the urine even when there is alkalosis is called "Paradoxical Aciduria".

4.How will you treat this patient ?
Answer-The patient has to be stabilised first by correcting nutritional status and metabolic abnormalities.

After stabilisation definitive surgery i.e. "Ramstedt's Pyloromyotomy" is performed.In the above surgery the serosa and muscularis is cut leaving the mucosa intact which relieves the obstruction.

Best Answer-Sanjeevani Hajra (SSKM and IPGMER Hospital)