Neuroblastoma Stage IV

Stage IV Neuroblastoma (metastatic NB) refers to high risk group of children with the primary tumor in the adrenal gland, mediastinum or pelvis associated with disease progression in other sites (bone marrow, cortical bone, liver, lymph node). Role of surgery in stage IV NB is controversial. Cure will require control of the primary tumor and elimination of metastatic disease. For infants with metastatic NB a more than 95% resection has been found adequate surgical treatment either initially or after effective chemotherapy. Adding ipsilateral lymph node dissection does not appear to affect survival. Delayed surgery after several courses of chemotherapy may be as effective as initial resection and is associated with fewer complications statistically. Resection without induction chemotox results in significant blood loss. High risk NB usually invades blood vessels and surrounding structures precluding resection. Intensive preop chemotherapy reduces tumor size and invasiveness allowing surgical removal. A fibrotic capsule forms with less blood supply to the tumor. Stage IV NB is best managed with initial chemotox until distant metastasis are controlled followed by primary gross tumor removal (even in the face of significant tumor reduction) and completion chemotox. Gross complete resection is best accomplished when a good partial response is obtained. Radiotox is added to unresectable lesions. Even when chemotox changes the tumor histology (Shimada) from unfavorable to favorable this does not improve overall outcome. Resection is not confounded by biology of the tumor (n-myc status). Survival is improved with kidney preservation during surgery. Local control of disease is a prerequisite for successful bone marrow transplantation.

References:
Incidental Appendectomy

Removing a normal appendix incidentally during a surgical procedure done for reasons other than abdominal pain is associated with a small but definite increase in adverse postoperative outcome. In this respect incidental appendectomy has been found to increase the incidence of postoperative septic complications (wound infection). It is neither cost-effective as an estimated 36 incidental procedures would be needed to prevent one case of appendicitis. As any procedure it increases adhesion formation from surgical manipulation in the right lower quadrant fossa. In potentially contaminated primary procedures the addition of incidental appendectomy does not increase operative morbidity or mortality. Incidental appendectomy is indicated in procedures where a potential diagnostic pitfall can occur such as Ladds procedure for malrotation, diagnostic laparoscopy for right quadrant pain and surgically reduced ileo-colic intussusception.

References:

Currarino Triad

Congenital caudal anomalies that include anorectal malformation, sacral bony abnormality and a presacral mass is known as the Currarino Triad (CT); an autosomic dominant hereditary syndrome described in 1981 caused by abnormal separation of neuroectoderm from endoderm. The anorectal malformation associated with CT is stenosis (or agenesis) of the distal rectum causing intractable constipation (chief complaint of this triad) or intestinal obstruction. Sacral agenesis and abnormalities of the os sacrum (scimitar sacrum, hemisacrum with preserved first sacral vertebra) are the most common bony anomalies identified. Most frequently the presacral mass in CT is reported to be an anterior meningocele, a benign teratoma, enteric, dermoid cyst or a combination. Though prenatal diagnosis can be made, most cases are diagnosed postnatally in the first decade of life. Routine pelvic x-rays should be done in all cases of anorectal stenosis. Pelvic ultrasound and x-rays in patients with history of chronic constipation since early childhood will suggest the diagnosis. MRI is the study of choice detecting the presacral mass and any anomalies of the spinal canal (tethered cord syndrome caused by the presacral mass). Management consists of excision of the presacral mass and repair of the anorectal
malformation. A gene associated with CT has been mapped to the terminal portion of the long arm of chromosome 7 (7q36).

References: