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## Completion Thyroidectomy

Thyroid nodules in children are managed based exclusively on the results of fine-needle aspiration (FNA) biopsy. Benign FNA results can be observed depending on the size, complexity, and symptomatology of the thyroid nodule. FNA results with positive pathology for papillary carcinoma is best managed with total thyroidectomy with central lymph node dissection. With results of FNA reported as indeterminate, suspicious, insufficient or a follicular neoplasm, removal of the affected lobe (hemithyroidectomy) might be performed. During removal of the affected lobe, the contralateral neck and remaining lobe should not be explored or violated. In cases in which a hemithyroidectomy is performed and the final pathology is reported as a well-differentiated papillary or follicular thyroid cancer, a completion thyroidectomy is performed. Completion thyroidectomy reduces locoregional recurrence, distant metastasis as well as low-risk carcinoma. The overall need for completion thyroidectomy in the era of FNA is generally less than 5-10% after lobectomy. The current major indications for completion thyroidectomy are gross extrathyroidal extension on the ipsilateral side, gross residual disease on the esophagus, recurrent laryngeal nerve, or the tracheal wall, major vascular or capsular invasion and poorly differentiated carcinoma or aggressive Hurthle cell carcinoma. Whenever a completion thyroidectomy is to be performed, the surgeon should study the gland previously removed to determine the status of the parathyroid glands that might be removed with the specimen. The rates of both temporary and permanent hypoparathyroidism are considerably higher in patients in whom the parathyroid glands were removed with the specimen. At the time of completion thyroidectomy when there is already a parathyroid gland in the initial hemithyroidectomy specimen, the surgeon must make every effort to identify and preserve both parathyroid and in the event of a suspected devascularization, the gland should be autotransplanted. Hemithyroidectomy followed by completion thyroidectomy does not appear to be associated with an increase operative risk of hypocalcemia or recurrent laryngeal nerve injury. The lower rate of temporary hypoparathyroidism and hypocalcemia seen in the completion thyroidectomy group can be attributed to the fact that the interval between operations allowed for recovery of any reversible injury caused at the initial hemithyroidectomy. Devascularized parathyroid glands require approximately four weeks to return to full function. Compared with total thyroidectomy, completion thyroidectomy has been associated with similar rates of recurrent laryngeal nerve injury and lower rates of hypoparathyroidism. After performing completion thyroidectomy, serum thyroglobulin levels tend to be an adequate prognostic follow-up marker. Beside the effectiveness of radioactive iodine for ablation of the remaining normal tissue or residual microscopic disease is enhanced after completion thyroidectomy. Recurrent laryngeal and superior laryngeal nerve

monitoring during completion thyroidectomy is associated with a decreased risk of injury. Completion thyroidectomy is a safe procedure with acceptable morbidity in the hand of experience surgeons.

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## Hematocolpos after Cloaca Repair

In females the most common anorectal malformation is an imperforate anus with a rectovestibular fistula, followed by rectoperineal fistula and the cloacal anomaly. Cloacal repair entails reconstruction of the urethra, vagina and rectum which end in a common channel. The aim of vaginal reconstruction is to provide a cosmetically satisfactory introitus, a conduit for normal menstruation and pain-free penetrative intercourse. There is a strong association of gynecologic anomalies (60%) associated with cloaca. A high rate of menstrual obstruction (40%) at puberty is well described. In less complex malformation such as rectovestibular and rectoperineal fistula a vaginal septum is the most common associated finding and can be managed most effectively during the initial repair of the rectum without trauma to the hymen or introitus. Vaginoscopy allows evaluation of the vaginal anatomy during infancy and puberty by documenting of vaginal duplication with two hemivaginas, and a septum, documentation of the septal length and total vaginal length. During vaginoscopy, perhaps at colostomy closure or creation of an appendicostomy, documentation of the presence of mucus at the outer part of the cervix can be evaluated. Also, cannulation of the distal fallopian tube with instillation of saline can be performed to visualize the egress of saline from the vagina and confirm patency of the Mullerian system. Hematocolpos is a medical condition of blood retained in the proximal vagina due to an outflow tract obstruction or blockage of menstrual flow. The most common cause of Hematocolpos in children without anorectal malformation is an imperforate hymen. In cases of repaired cloaca, a longitudinal or transverse vaginal septum, congenital or acquired vaginal atresia or severe vaginal stricture, uterus didelphis and septate uterus can cause Hematocolpos. Ovarian function is normal in girls with repaired cloaca so pubertal and breast development occurs as expected. Thelarche (breast development) occurs between 9-10 years of age with menstruation occurring at 12-12.5 years of age. Confirmation of the patency of the reproductive tract before menarche is important to avoid obstruction, pain, and risk to reproductive organs with infertility. It is during this time that repaired cloaca can be studied further to determine if there could exist the possibility of menstrual flow tract

obstruction and development of hematometrocolpos by performing serial ultrasound studies. Almost 20% of children born with cloaca develop amenorrhea due to absence of or underdeveloped Mullerian structures. US surveillance of the reproductive structures should begin 6-9 months after thelarche and continue every six months through menarche. If an obstruction to menstrual flow is detected by visualization of a thickened endometrium with hematocolpos, medical intervention should be initiated immediately to minimize adverse sequelae. Hormonal suppression of menses and endometrial stimulation should be started if there is menstrual flow obstruction to prevent continued accumulation of blood. Surgery is usually necessary to establish an adequate outflow tract either by resection of a vaginal septum, introitoplasty, posterior vaginoplasty or vaginal replacement with bowel in cases of absent vagina. Other gynecologic concern for pubertal females includes the development of adnexal cysts, hydrosalpinxes, endometriosis and chronic pelvic pain. Women who had a history of a cloacal anomaly should be delivered by cesarean section.

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## Apple Peel Atresia

Jejunoileal atresias are classified as type I characterized by a transluminal septum; type II involves a fibrous cord connecting two blind ending pouches; type IIIA has a V-shaped mesenteric defect; and type IV exhibits multiple atretic segments. The term Apple peel atresia (or Type IIIB intestinal atresia), occurring in less than 10% of all jejunoileal atresias, refers to the development of a high jejunal atresia with discontinuity of the small bowel and a wide gap in the mesentery. The distal segment of jejuno-ileum is shortened and assumes a helical configuration around a retrograde perfusing vessel. The appearance is like a Christmas tree, hence the synonym Christmas tree deformity. An intrauterine vascular accident after emergence of the middle colic artery to the affected proximal bowel during late gestation has been accepted as the cause of apple-peel atresia presenting with a wide spectrum of occlusions of one or more branches of the superior mesenteric artery. The bowel distal to the atresia is precariously supplied in a retrograde fashion by anastomotic arcades from the ileocolic, right colic or inferior mesenteric artery. Most of these children have less than half of the normal length of the small bowel and a physiological short bowel. Apple peel atresia is usually reported as an isolated malformation, but has also been related to malrotation, situs inversus and polysplenia. Not all distal apple-peel atresias are associated with a proximal bowel atresia since a few scattered reports of a mesenteric

defect associated with a marginal artery may cause the coiling defect of the apple peel as the bowel outgrows it blood supply causing problems of ischemia later in life associated with mesenteric internal hernias. Also, not all apple-peel atresias are from the proximal jejunum, since very few have been described arising after a proximal duodenal atresia related to the second portion of the duodenum with absence of the third and fourth portions of duodenum and superior mesenteric artery. Infants born with high jejunal atresia have considerable dilatation of the proximal bowel, while the distal segment is small and collapsed. Anastomosis between two discrepant bowel sizes can cause functional bowel obstruction. The resulting peristalsis is incapable of producing an adequate upstream pressure gradient. As alternative, an antimesenteric reduction-tapering proximal jejunoplasty can be used to perform anastomosis reducing the caliber of the proximal bowel to fit an almost end to end anastomosis with the distal microbowel. Serial transverse enteroplasty of the dilated proximal jejunal atresia can also be performed to reduce the caliber while lengthening it appropriately without significant loss of absorptive area. Apple-peel atresias frequently have a high incidence of prematurity, short gut, multiple atresias and associated anomalies which constitute potential prognostic factors. Antenatal US may suggest the diagnosis of jejunoileal atresia with the presence of dilated fluid-filled loops of bowel and polyhydramnios. Surgery is the preferred mode of treatment for jejunoileal atresias and the goal of treatment is to establish bowel continuity while preserving as much bowel length as possible. Postoperative complications associated with apple-peel atresia include anastomotic dysfunction (most common), sepsis (from leak or TPN), short bowel syndrome, necrotizing bowel, stenosis and even death. Survival is above 90%.

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