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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 chemotx 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 chemotx until distant metastasis are controlled followed by primary gross tumor removal (even in the face of significant tumor reduction) and completion chemotx. Gross complete resection is best accomplished when a good partial response is obtained. Radiotx is added to unresectable lesions. Even when chemotx 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:

1- DeCou JM, Bowman LC, Rao BN, Santana VM, Furman WL, Luo X, Lobe TE, Kumar M: Infants with metastatic neuroblastoma have improved survival with resection of the primary tumor. J Pediatr Surg 30(7): 937-941, 1995

2- La Quaglia MP, Kushner BH, Heller G, Bonilla MA, Lindsley KL, Cheung NKV: Stage 4 neuroblastoma diagnosed at more than 1 year of age: Gross total resection and clinical outcome. J Pediatr Surg 29(8): 1162-1166, 1994

3- Shamberger RC, Allarde-Segundo A, Kozakewich HPW, Grier HE: Surgical management of stage III and IV neuroblastoma: Resection before or after chemotherapy? J Pediatr Surg 26(9): 1113-1118, 1991

4- Haase GM, O'Leary MCO, Ramsay NKC, Romansky SG, Stram DO, Seeger RC, Hammond GD: Aggressive surgery combined with intensive chemotherapy improves survival in poor-risk neuroblastoma. J Pediatr Surg 26(9): 1119-1124

5- Shorter NA, Davidoff AM, Evans AE, Ross AJ 3rd, Zeigler MM, O'Neill,JA Jr: The role of surgery in the management of stage IV neuroblastoma: a single institution study. Med Pediatr Oncol 24(5):287-91, 1995

6- Tsuchida Y, Yokoyama J, Kaneko M, Uchino J, Iwafuchi M, Makino S, Matsuyama S, Takahashi H, Okabe I, Hashizume K, et al: Therapeutic significance of surgery in advanced neuroblastoma: a report from the study group of Japan. J Pediatr Surg 27(5):616-22, 1992

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:
1- Wen SW, Hernandez R, Naylor CD: Pitfalls in non-randomized outcomes studies. The case of incidental appendectomy with open cholecystectomy. JAMA 274(21):1687-91, 1995
2- Miranda R, Johnston AD, O'Leary JP: Incidental appendectomy: frequency of pathologic abnormalities. Am Surg 46(6):355-7, 1980
3- Mulvihill S, Goldthorn J, Woolley MM: Incidental appendectomy in infants and children. Risk v rationale. Arch Surg 118(6):714-6, 1983
4- Sugimoto T, Edwards D: Incidence and costs of incidental appendectomy as a preventive measure. Am J Public Health 77(4):471-5, 1987
5- Addiss DG, Shaffer N, Fowler BS, Tauxe RV: The epidemiology of appendicitis and appendectomy in the United States. Am J Epidemiol 132(5):910-25, 1990
6- Ritchey M, Haase GM, Shochat SJ, Kelalis PP: Incidental appendectomy during nephrectomy for Wilms' tumor. Surg Gynecol Obstet 176(5):423-6, 1993

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:

1- Kirks DR, Merten DF, Filston HC, Oakes WJ: The Currarino triad: complex of anorectal malformation, sacral bony abnormality, and presacral mass. Pediatr Radiol 14(4):220-5, 1984
2- Kochling J, Pistor G, Marzhauser Brands S, Nasir R, Lanksch WR: The Currarino syndrome--hereditary transmitted syndrome of anorectal, sacral and presacral anomalies. Case report and review of the literature. Eur J Pediatr Surg 6(2):114-9, 1996
3- Lee SC, Chun YS, Jung SE, Park KW, Kim WK: Currarino triad: Anorectal malformations, sacral bony abnormality, and presacral mass - A review of 11 cases. J Pediatr Surg 32(1): 58-61, 1997
4- Gudinchet F, Maeder P, Laurent T, Meyrat B, Schnyder P: Magnetic resonance detection of myelodysplasia in children with Currarino triad. Pediatr Radiol 27(12):903-7, 1997
5- Zia-ul-Miraj M, Brereton RJ: Currarino's triad: an unusual cause of constipation in children. Pediatr Surg Int 13(5-6):437-9, 1998
6- Samuel M, Hosie G, Holmes K: Currarino triad-Diagnostic dilemma and a combined surgical approach. J Pediatr Surg 35(12):1790-1794, 2000

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