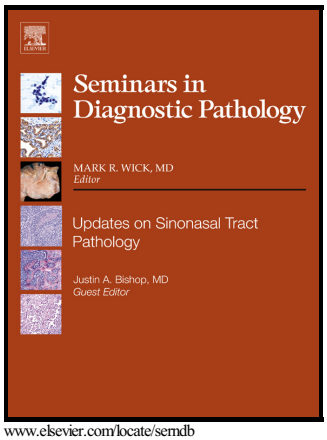
Testicular germ cell tumors (TGCT) in neonates and infants

Juan Carlos Manivel



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Testicular Germ Cell Tumors (TGCT) in neonates and infants

Juan Carlos Manivel, M.D.

Department of Pathology and Laboratory Medicine.

Veterans Administration Medical Center

One, Veterans Drive. BB-113

Minneapolis, MN 55417

Introduction:

What occurs to "pediatric" patients is influenced by biologic changes that occur during development and adolescence and the different susceptibility during this period to internal and external factors that can cause disease. Specialists of various medical disciplines involved in the treatment of "pediatric" patients usually include in their practice patients in the first two decades of life. In the field of TGCT two very different types of neoplasms occur in "pediatric" patients: the very young, and the adolescent. TGCT are a heterogeneous group of neoplasms; the definition terminology and classification used here follow the 2016 WHO classification of TCGT (Table 1) (Ref 1). Criteria employed in this classification include whether or not the tumor is invasive, whether it is of a single or more than one histological type, whether it occurs in pre- or post-pubertal gonads, and whether or not it is related to germ cell neoplasia in-situ (GCNIS, formerly intratubular germ cell neoplasia, unclassified/carcinoma in-situ). On the basis of their biology and clinical features, germ cell tumors are classified into three types that correlate with specific ages at presentation. The TGCT in type I are the subject of this review, and include teratomas and yolk sac tumors (YST, endodermal sinus tumors) that occur in neonates and infants; well before puberty as most cases occur in the four first years of life. In this review they will be referred to as prepubertal teratomas and YST. Incidentally, type I also includes teratomas and YST in the ovary, sacrococcygeal and retroperitoneal regions, the head and neck, the pineal and hypothalamic-hypophyseal region, and the vagina (Ref 2). Type II represented by embryonal carcinoma, seminoma, and mixed germ cell tumors, is the most numerous and includes those tumors that present during adolescence and young adulthood; they will be referred to here as postpubertal TGCT. Some of these tumors occurring in adolescents have mistakenly been included in the past among the "pediatric" (type I) cases. However, type II tumors essentially never occur before 5 years of age, while most type I tumors occur before this age. In about 5 % of patients with type II tumors, these occur in extragonadal sites. Type III includes the spermatocytic tumor (formerly spermatocytic seminoma) that occurs in elderly men. Types II and III do not occur in the infant testicle. Although the focus of this essay is on type I TGCT, relevant aspects of Types II and III will be discussed to contrast differences in relation to those in type I.

The annual incidence of prepubertal TCGT (0.12 per 100,000) is considerably lower than in adults (6 per 100,000) and lower than that of spermatocytic tumors in older adults (0.2 per 100,000). Annual increase