Staging

In patients with a malignant tumour (yolk sac tumour, immature teratoma) staging should be performed including either a CT-scan or MRI of the abdomen and a CT-scan of the chest. If there is any suspicion of a non-organ confined tumour, the patient should be referred to a paediatric oncologist. In patients with the rare diagnosis of a Granulosa cell tumour, imaging of the abdomen to exclude enlarged lymph nodes is reasonable as this may be a potentially malignant tumour; in those with Sertoli or a Leydig cell tumour, an MRI is recommended, as 10% are malignant and the metastases do not respond very well to chemotherapy or radiation in the adult literature [173, 174]. For information about staging of testicular tumours, we refer to the EAU Testicular Cancer Guidelines [138]. In benign tumours (mature teratoma, epidermoid cysts) no further staging is required.

*3.3.4 **Treatment/Management***

If a testicular tumour is suspected, surgery with the option of intra-operative frozen section should be performed. It is not necessary to do this as an emergency procedure. However, to confirm the diagnosis and to avoid familial anxiety, the operation should be scheduled as soon as possible, preferably within the next few days. Testis-sparing surgery (TSS) should be performed, whenever possible. In a systematic review, the recurrence rate after surgery was found to be 5.8% (95% CI: 2.3-14.1%) in cohorts including a benign rate of 70.9% [175]. Testis-sparing surgery was performed in 36.2% of these patients.

When performing TSS, frozen sections during surgery could be performed to confirm the diagnosis (benign vs. malignant tumour) and to confirm if a microscopically margin-negative resection is performed, in which no gross or microscopic tumour remains in the primary tumour bed (R0 resection). In cases of an R0 resection, the tunica is closed and the testis is replaced in the scrotum. In case of R1 resection (removal of all macroscopic disease, but microscopic margins are positive for tumour) confirmed by frozen section in a malignant or potential malignant tumour, an orchiectomy should be performed at the same time of surgery. If the final pathology later demonstrates a R1 resection in a malignant tumour despite intra-operative negative margins on frozen section, an inguinal orchiectomy can safely be performed. Similarly, in a multi-centre retrospective study, intra-operative biopsy followed by tumorectomy to preserve healthy testicular tissue in benign testicular tumours was recommended [176].

Orchiectomy could be considered only if normal testicular parenchyma is no longer detectable in the pre-operatively high-resolution US and/or the AFP is > 100 ng/mL in a > 12-month-old boy, which is highly suspicious of a yolk sac tumour.

For surgical technique, the Panel is in favour of an inguinal approach. Furthermore, clamping of the vessels has the advantage of a better view, when organ sparing surgery is performed. However, there is no evidence in the literature that tumour-spread is prevented by clamping the vessels. In addition, a retrospective multicenter study suggested that a scrotal approach may be applied for selected cases [177].

As most paratesticular tumours are benign, intra-operative frozen section should be available during surgery and organ sparing surgical approach is preferred in benign tumours. If a paratesticular rhabdomyosarcoma is suspected, radical inguinal orchiectomy should be performed if tumour size allows it, otherwise it is better to extend the inguinal incision down to the scrotum or use a combined inguinal and scrotal approach to facilitate a complete gross total tumour resection [178].

*3.3.5 **Tumour entities in prepubertal boys***

3.3.5.1 Germ cell tumours

Teratomas are usually benign in prepubertal children and represent the greatest proportion of intratesticular tumours (around 40%) [149, 179]. They present at a median age of thirteen months (0-18 months). Only in adolescents and adults, they should be considered as malignant tumours. Histologically they can consist of a combination of the three primitive embryological germ-cell layers (ectoderm, mesoderm and endoderm). Most of these elements shows microscopically mature elements [180]; however, some immature teratomas in this age group have also been reported [181]. On US examination a heterogenous picture with some calcification is seen [182] and AFP should be less than 100 ng/mL in an infant. After organ-sparing surgery only one recurrence was reported in the literature [183].

Epidermoid cysts are of ectodermal origin and seem to be related to well-differentiated teratomas; they are always benign [180]. Keratin-producing epithelium is responsible for the keratinised-squamous-epithelial deposits, which appear hyperechogenic in an US [182]. Organ-sparing surgery should be performed and if confirmed by histology, there is no need for surveillance despite the fact that one “recurrence” has been reported thirteen years after diagnosis [184].