



Review article

Ovarian cancer in children and adolescents: A rare disease that needs more attention



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ABSTRACT

Ovarian cancer is rare in childhood. This explains why there are only scattered reports on it in the literature and why there is a lack of specific pediatric treatment. This paper gives an overview of the Belgian data from 2004 to 2013 and reviews the literature. To index ovarian masses and malignancies in children better in the future, worldwide data collection should be improved and reproducible definitions of 'childhood', 'malignancy' and 'ovarian mass' need to be adopted.

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1. Ovarian cancer in children: a rare disease

The most important childhood cancers are: leukemia, central nervous system tumors and neuroblastoma [1]. Tumors of the female genital tract in children and adolescents are rare. Amongst them, ovarian cancer is the most frequent, accounting for 1% of

all childhood cancers [2]. According to the SEER database (the Surveillance, Epidemiology and End Results of the National Cancer Institute, USA), 1.3% of all ovarian cancers between 2008 and 2012 were diagnosed in patients below the age of 20. This makes ovarian cancer in children extremely rare and therefore hard to study. Benign ovarian masses on the other hand are more frequent, affecting approximately 2.6/100 000 girls each year [3].

The distribution of pathology in ovarian cancer is different in children than it is in adults (Table 1). Ovarian cancer is the most important gynaecological cancer in children, but a far second in adult women [4]. Epithelial ovarian carcinomas account for 90%

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Table 1
Classification of ovarian tumors according to the WHO classification.

Epithelial	Serous	
	Mucinous	
	Endometrioid	
	Clear cell	
	Transitional cell	
Non-epithelial	Epithelial-stromal (adenosarcoma, carcinosarcoma)	
	Germ cell	Dysgerminoma
		Yolk sac
		Monodermal
		Mixed
		Teratoma
	Sex cord stromal	Granulosa cell tumor
		Sertoli-Leydig
		Mixed
	Metastases	

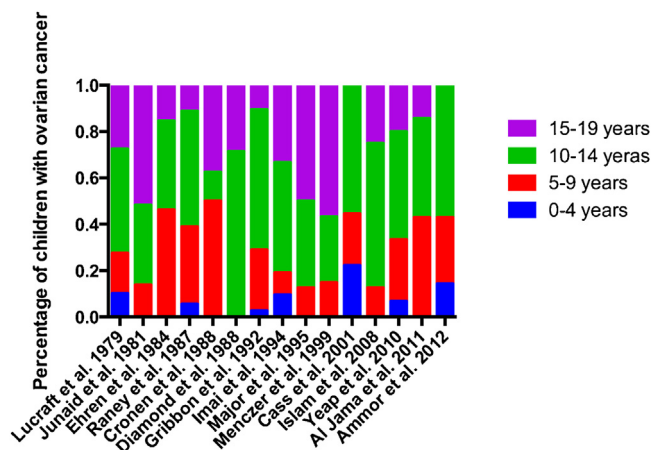


Fig. 1. Age distribution of ovarian cancer cases in children in literature. Absolute numbers are shown. [8,10–41].

of all ovarian cancers in adults. High-grade serous ovarian cancer (HGSOC) is the most frequent subtype. The prognosis of HGSOC remains poor. This explains the poor prognosis of ovarian cancer in adults. HGSOC metastasizes throughout the abdomen before causing symptoms, resulting in diagnostic delay and leading to a poor prognosis. Nowadays, 80% of adult ovarian cancer patients still die. In children, however, the most frequent ovarian cancer is non-epithelial. Most often, their ovarian cancers arise from germ cells. Less than 20% of ovarian cancers in children are epithelial in origin, with a predominance of serous and mucinous histology [5–7]. The overall prognosis of ovarian cancer in children is excellent compared with that in adults, because of their different histological distribution [7,8]. According to a study by Mangili et al. reporting results from the largest database on ovarian germ cell pathologies, there are two important contributing factors to the prognosis of GCT: first, the majority (71%) is detected at stage I; and second, they respond well to surgery and chemotherapy, leading to a 5-year survival of 95.6% and 73.2% in stage I and advanced stages, respectively [9]. Because of this excellent prognosis of germ cell tumors (GCTs), overall outcome of ovarian cancer in children is excellent compared with that in adults.

2. Literature review

Table 2 and Fig. 1 give a summary of the cases of ovarian childhood cancer reported in the literature [8,10–41]. It is clear that this represents a scattered overview of pediatric ovarian neoplasms, which is difficult to interpret. Each center publishes locally available data. The number of ovarian childhood cancers as a proportion

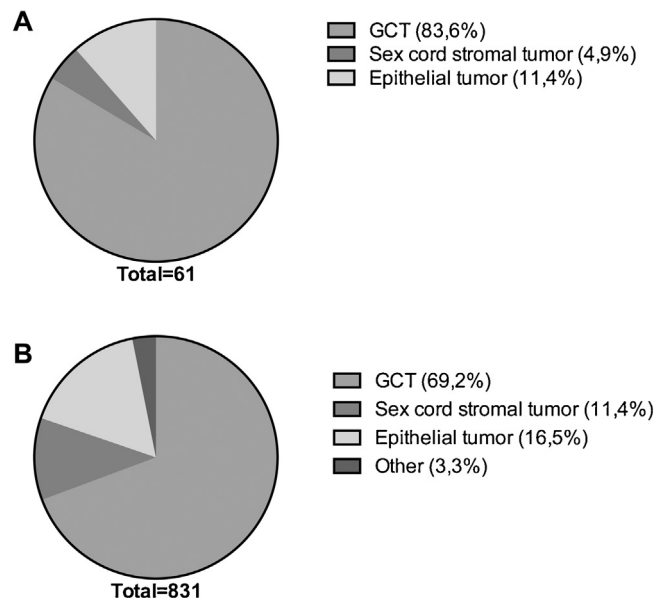


Fig. 2. Distribution of different histological types of ovarian cancer derived from the Belgian data (A), literature (B) [8,19–23,30–45,47–52]. Germ cell tumors are most frequent in both series, followed by epithelial tumors.

of the total number of ovarian masses (both benign and malignant) differs enormously. On average, this results in a risk of malignancy of adnexal masses in children of around 19%, with a range from 2 to 59%. This is an enormous variation. This difference cannot be explained by geographical differences alone. For this we see three additional causes. (1) Benign cysts and teratomas are often treated locally and not referred to a specialized pediatric oncologist and therefore will remain underreported. This artificially lowers the total number of ovarian masses seen and artificially increases the proportion of cancers. (2) There are differences in the study populations used in the case series. Some groups only report adnexal masses up to the age of 15, as was done by Del Mar et al., while other groups include patients until the age of 19 [8]. Children with cancer up to the age of 16 years are referred to a pediatric oncology unit. (3) The terms and definitions used to describe ovarian masses differ from paper to paper. The gold standard for the diagnosis of an ovarian neoplasm is the ultrasound definition provided by the IOTA (International Ovarian Tumor Analysis) criteria: it is the part of an ovary or the total ovary that is not part of the normal ovarian physiology [42].

Despite differences in reporting, it is clear from the Belgian data, as from the data reported more generally in the literature, that there is a positive correlation of ovarian cancer with age (Fig. 2)

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