Ovarian or paraovarian lesions in children

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EVIDENCE BASE

1. Ovarian/paraovarian masses (including cysts) are infrequent in childhood but may be encountered at any age, either incidentally, electively or as an emergency. Management should be based on the principle of "first do no harm".

2. The majority of cystic ovarian lesions are benign, BUT NOT ALL.

3. Solid masses are usually tumours which may be benign or malignant.

4. Expert ultrasound is the key to successful diagnosis and optimal management, and should be undertaken preoperatively whenever possible.

5. Any patient with a possible ovarian tumour (solid or cystic) should have tumour markers assessed preoperatively.

6. Ovarian sparing and protection of fertility is a key aspect of all ovarian surgery and should be prioritised at all times except if it will result in potential "upstaging" or spread of a malignancy.

7. Gynaecologists are 8-15 times more likely to preserve the ovary than paediatric general surgeons therefore gynaecology input/advice should be sought before surgery in all cases.

8. Even though most paediatric ovarian malignancies are low-grade and responsive to treatment, all efforts must be made to avoid "upstaging" a malignancy through inadvertent or deliberate rupture of the cyst or mass during diagnosis or removal (including aspiration of cysts).

9. Laparoscopic-assisted surgical removal of masses or large cysts is feasible, but all potentially malignant masses or cysts should be removed intact through a separate incision to avoid rupture, seeding or upstaging of any tumour.

10. Surgical staging (whether open or laparoscopic) is a key part of the assessment of any ovarian malignancy.

11. Cytology of any fluid removed and full pathological examination of all excised tissue is mandatory

All the above statements are based on published evidence from the last 5 years.

PRINCIPLES OF MANAGEMENT

1. "Do no harm" i.e.:
    - Avoid unnecessary surgery
    - Ensure ovarian sparing whenever possible
    - Ensure malignancy potential is assessed fully
    - Avoid "upstaging" any tumour

2. All paediatric surgeons of any grade undertaking elective or emergency abdominal surgery should be aware of
   - the risk factors for malignancy in ovarian masses
   - the need for ovarian-sparing surgery
   - the need to follow these guidelines for ovarian lesions

3. Experts should be involved at all stages
   - Consultant Paediatric Surgeons to be involved directly in all cases
   - Discuss all cases (except conservative management of simple neonatal ovarian cysts) with gynaecologists - before surgery, +/-intraoperatively and in planning follow-up
   - Expert cytology/pathology to be requested on all specimens

4. The guidelines should followed according to the age of the patient and regardless of whether the lesion is encountered electively, incidentally, or as an emergency.

5. If an ovarian cyst/mass is encountered unexpectedly at any paediatric surgical operation (emergency or elective) it is better to leave the lesion complete and in situ (after detorsion if necessary) until the necessary radiological workup and marker studies have been done and gynaecological opinion has been sought, even if this means a second procedure is required.

6. Individual cases and all results should be reviewed by a Multi-disciplinary team (including paediatric surgery, gynaecology, oncology, pathology and radiology) on a regular basis (*interval to be agreed depending on anticipated numbers of cases*).

7. Regular audit of diagnoses, management, outcomes and adherence to guidelines should be performed (*frequency will be dependent on caseload, but should be at least every 2 years and preferably annual?*).

8. It is advisable to use a "checklist" for the management of patients with ovarian lesions to enhance education and facilitate future audit.

Useful tumour markers	AFP: Germ cell and Sertoli-Leydig tumours β-hCG, LDH: Germ cell tumours CA-125: Epithelial tumours and endometrioma *Note:* Immature malignant teratomas do not produce elevated markers
Characteristics of malignancy	Lesion >10cm, complex structure on US, nodular or papillary appearance, thick septations, ascites, peritoneal seeding, lymphadenopathy, hepatic metastases "Malignancy scoring index" $\geq$7
"Malignancy scoring index"	MSI = Total of the following: Maximum diameter of solid component in cm + presence/absence of sex-hormone-related symptoms (score 6 if present, 0 if absent) + enhancement or flow in a septum or solid papillary projection (score 4 if present, 0 if absent) Total $\geq$7 is highly likely to be malignant lesion (sensitivity 87.5-100%, sensitivity 97.9-100%)

MANAGEMENT GUIDELINES A

ELECTIVE SITUATION

AGE ACTION	Neonate (pre- or postnatal diagnosis)	1m-1year	1-10 years	>10 years
Detailed USS of whole abdomen	Yes	Yes	Yes	Yes
Tumour markers	Yes except for simple unilocular cysts	Yes	Yes	Yes
Other imaging indicated? (eg Chest & abdomen Xray, CT or MRI)	Yes if malignancy suspected or possible from US findings	Yes if malignancy suspected or possible from US findings	Yes if malignancy suspected or possible from US findings	Yes if malignancy suspected or possible from US findings
Gynaecology opinion Required	Yes if ovarian sparing may be required	Yes if ovarian sparing may be required	Yes	Yes
Conservative management indicated?	Yes – for simple unilocular cysts provided under regular US surveillance, no symptoms, and no risk of torsion or malignancy	Yes – for simple unilocular cysts provided under regular US surveillance, no symptoms and no risk of torsion and no risk of malignancy	Yes – but only according to gynaecology advice +/- participation	Yes – but only according to gynaecology advice +/- participation
Laparoscopic surgery indicated?	Only for aspiration of simple unilocular cysts with no resection required	i) Yes – for aspiration of simple cysts ii) Laparoscopic-assisted removal of complex or solid lesions may be possible - lesion should be removed intact with maximum ovarian sparing	Only after opinion +/- participation of gynaecologist. Lesion should be removed intact with maximum ovarian sparing	Only after opinion +/- participation of gynaecologist. Lesion should be removed intact with maximum ovarian sparing
Open surgery indicated?	Yes for large or complex lesions: Ensure intact removal + maximum safe ovarian sparing + macroscopic staging +/- LN sampling	Yes for large or complex lesions: Ensure intact removal + maximum safe ovarian sparing + macroscopic staging +/- LN sampling	Yes for large or complex lesions: +/- gynaecologist: + intact removal + maximum safe ovarian sparing + macroscopic staging +/- LN sampling	Yes for large or complex lesions: + gynaecologist + intact removal + maximum safe ovarian sparing + macroscopic staging +/- LN sampling
Follow-up	Paediatric Surgery	Paediatric Surgery	Paediatric Surgery +/- Gynaecology	Gynaecology +/- Paediatric Surgery

MANAGEMENT GUIDELINES B

EMERGENCY SITUATION

If an ovarian / paraovarian pathology is a possibility in a patient presenting as an emergency (e.g. adolescent girls with acute abdominal pain) the consultant on call MUST be informed, and a preoperative US should be done. If an ovarian / paraovarian lesion is confirmed, the ELECTIVE guidelines should be followed. **The following guidelines refer to the unexpected finding of an ovarian / paraovarian lesion at the time of surgery, whether the surgery is elective, emergency, laparoscopic or open.**

AGE	Neonate	1m-1year	1-10 years	>10 years
Lesion palpable pre-op	Inform consultant, request advice +/- presence. Delay surgery, do US and take blood for tumour markers. Follow elective guidelines	Inform consultant, request advice +/- presence. Delay surgery, do US and take blood for tumour markers. Follow elective guidelines	Inform consultant, request advice +/- presence. Delay surgery if feasible, do US and take blood for tumour markers. Follow elective guidelines	Inform consultant, request advice +/- presence. Delay surgery if feasible, do US and take blood for tumour markers. Follow elective guidelines.
Non-torted lesion found incidentally at laparoscopic procedure.	Inform consultant. No surgical action except aspiration of simple cyst. Request post-op US and start discussion with parents about further management. Follow elective guidelines	Inform consultant. No surgical action except aspiration of simple cyst. Take blood for tumour markers. Request post-op US and start discussion with parents about further management. Follow elective guidelines	Inform consultant. No surgical action. Take blood for tumour markers. Leave ovarian lesion intact. If LN enlarged, take biopsy. Request post-op US and follow elective guidelines	Inform consultant. No surgical action. Take blood for tumour markers. Leave ovarian lesion intact. If LN enlarged, take biopsy. Request post-op US and follow elective guidelines
Non-torted lesion found incidentally at open surgery for other emergency	Inform consultant. If simple cyst, aspirate. If large, complex or solid lesion, excise INTACT maximum ovarian sparing and check for enlarged LN. Inform parents and follow elective guidelines.	Inform consultant. If simple cyst, aspirate. If large, complex or solid lesion, take blood for tumour markers, excise INTACT with maximum ovarian sparing and check for enlarged LN. Inform parents and follow elective guidelines.	Inform consultant. Take blood for tumour markers. Consider need for gynaecology advice. If large, complex or solid lesion, excise INTACT with maximum ovarian sparing and check for enlarged LN. Inform parents and follow elective guidelines	Inform consultant. Take blood for tumour markers. Request gynaecology assistance. Evaluate lesion and check for enlarged LN: if present, take LN biopsy. Do not breach ovarian lesion. No further action indicated. Inform parents and follow elective guidelines
Torted or haemorraghic lesion found unexpectedly at laparoscopic or open procedure for other diagnosis	Inform consultant and request presence. Take blood for tumour markers. Detort lesion. If single cyst, aspirate. If solid or infarcted, excise INTACT with maximum ovarian and adnexal sparing. Finish procedure and follow elective guidelines.	Inform consultant and request advice. Take blood for tumour markers. Detort lesion. If solid or infarcted, excise INTACT with maximum ovarian and adnexal sparing. Finish procedure and follow elective guidelines.	Inform consultant and request presence. Take blood for tumour markers. Consider need for gynaecology advice. Detort lesion. If solid or infarcted, excise INTACT with maximum ovarian and adnexal sparing. If cystic, leave in situ. Follow elective guidelines.	Inform consultant and request presence. Take blood for tumour markers. Request gynaecology advice +/- assistance. Detort lesion. If solid or infarcted, excise INTACT with maximum ovarian and adnexal sparing. If cystic, leave in situ. Follow elective guidelines.