BACKGROUND

Key recommendations

- Craniopharyngiomas are rare benign central nervous system tumours arising from embryological remnants of Rathke’s pouch, but account for 80% of all suprasellar lesions in children and young people (CYP).
- 30-year overall survival rates are high (up to 80%) but patients often suffer multiple relapses requiring repeated interventions and long-term neuroendocrine, cognitive and visual-tumour- and treatment-related morbidity.
- Treatment is usually by a combination of neurosurgery and/or radiotherapy but the rarity of these tumours and the lack of randomised-controlled trials means that there are large variations in management between centres with no previous evidence-based national or international guidelines.
- To provide, for the first time, an evidence- and consensus-based standard for best practice for the diagnosis, assessment, management and follow-up of craniopharyngiomas in CYP aged <19 years under the auspices of the Royal College of Paediatrics & Child Health (RCPCH), Children’s Cancer & Leukaemia Group (CCLG) and the British Society for Paediatric Endocrinology & Diabetes (BSPED).

METHODS


3-tier exclusion filter: Tier 1: Excluded based on title Tier 2: Excluded based on abstract Tier 3: Excluded based on detailed reading of full paper

GRADE assessment of quality of evidence

Consensus recommendations made based on GDG Development Group expert opinion for statements where no/insufficient published evidence was obtained

32 statements of recommendation

33 consensus recommendations

28 consensus recommendations included (total 40 recommendations made)

AIM

To provide, for the first time, an evidence- and consensus-based standard for best practice for the diagnosis, assessment, management and follow-up of craniopharyngiomas in CYP aged <19 years under the auspices of the Royal College of Paediatrics & Child Health (RCPCH), Children’s Cancer & Leukaemia Group (CCLG) and the British Society for Paediatric Endocrinology & Diabetes (BSPED).

Conclusions

These UK RCPCH/ CCLG/ BSPED-endorsed guidelines provide the first evidence-based recommendations for the management of paediatric craniopharyngiomas, by which better consistency in the quality of care of such patients may be achieved with the aim of improving long-term quality of survival.