



Multidisciplinary management and long-term outcomes in paediatric craniopharyngioma: An 8-year single-centre experience

Authors

**Dr. Naga Bhagyasri Mangam¹, Dr. Prasun Deb², Dr. Smitha Nalla³,
Dr. Sandeep Devireddy⁴, Dr. Manas Panigrahi⁵, Dr. M Sailaja⁶, Dr. I Satish Rao⁷**

¹MBBS, MD, DrNB- Resident in Endocrinology, Krishna Institute of Medical Sciences Hospital, Secunderabad

²MBBS, MD, DM – HOD Department of Endocrinology; Krishna Institute of Medical Sciences Hospital, Secunderabad

³MBBS, MRCP, CCT, FRCP(Edin), Consultant Endocrinologist; Krishna Institute of Medical Sciences Hospital, Secunderabad

⁴MBBS, DNB, DM, Consultant Endocrinologist; Krishna Institute of Medical Sciences Hospital, Secunderabad

⁵MBBS, MS, MCh, FACS, HOD- Department of Neurosurgery; Krishna Institute of Medical Sciences Hospital, Secunderabad

⁶MBBS, MD, HOD- Department of Pathology; Krishna Institute of Medical Sciences Hospital, Secunderabad

⁷MBBS, MD Pathology, Senior Consultant Pathologist, Krishna Institute of Medical Sciences Hospital, Secunderabad

Abstract

Background: Craniopharyngiomas are benign epithelial tumors derived from remnants of Rathke's pouch, typically arising in the sellar and parasellar regions. Despite their benign histology, they demonstrate locally aggressive behaviour; often involving critical neurovascular structures such as the hypothalamus. Management is challenging, and long-term morbidity is common.

Methods: We retrospectively analysed 10 paediatric patients (≤ 18 years) with histologically confirmed craniopharyngioma treated in the Department of Endocrinology, Krishna Institute of Medical Sciences, over 8 years, focusing on clinical presentation, treatment strategies, and long-term endocrine and metabolic outcomes.

Results: The median age at diagnosis was 12 years (range 3–18 years); 6 patients were boys. Presenting symptoms included headache ($n=10$), vomiting ($n=4$), visual impairment ($n=2$), seizures ($n=1$), motor weakness ($n=1$), irregular menstrual cycles ($n=1$), and absent secondary sexual characteristics ($n=1$). Pre-operative interventions included ventriculoperitoneal shunt ($n=4$), endoscopic cyst aspiration ($n=1$), and Ommaya reservoir placement ($n=2$). All patients underwent surgical resection, followed by adjuvant radiotherapy (8 conventional, 1 stereotactic, 1 proton beam). Post-treatment hormone replacement therapy was required in most patients, primarily glucocorticoids ($n=8$) and levothyroxine ($n=7$). At follow-up, 4 patients were obese, 4 overweight, 1 had impaired glucose tolerance, 1 had diabetes mellitus, 1 had hypertension, and 1 had dyslipidaemia. Seven patients remain progression-free (median follow-up: 8 years); three patients died, two due to recurrence.

Conclusion: Paediatric craniopharyngiomas demonstrate excellent overall survival but high rates of long-term endocrine insufficiency and metabolic syndrome, especially with hypothalamic involvement. Long-term follow-up focusing on cardiovascular risk factor screening and optimal hormone replacement therapy is essential for improving quality of life and outcomes.

Keywords: Craniopharyngioma, paediatric brain tumour, Rathke's pouch, panhypopituitarism, metabolic syndrome, proton therapy.

Introduction

Craniopharyngiomas are rare, benign, epithelial tumours that arise from remnants of Rathke’s pouch in the sellar or parasellar regions. Despite their benign histopathology, they are locally aggressive and frequently involve the optic apparatus, pituitary gland, and hypothalamus, complicating management and long-term outcomes.

The incidence is estimated at 0.5–2 cases per million population per year, accounting for 1.2–4% of paediatric intracranial tumours. Standard treatment modalities include surgical resection and/or radiotherapy. However, due to their proximity to vital neural structures, complete resection is often not possible without significant morbidity.

Long-term complications in survivors are common and include panhypopituitarism, diabetes insipidus, obesity, and metabolic syndrome. These complications can adversely affect quality of life and long-term survival. Here, we present an 8-year retrospective analysis of 10 paediatric patients with histologically confirmed craniopharyngioma managed in the department of Endocrinology, Krishna Institute of Medical Sciences, focusing on clinical presentation, management strategies, and long-term endocrine/metabolic outcomes.

Methods

We performed a retrospective review of medical records of paediatric patients (≤18 years) with histologically proven craniopharyngioma treated at our centre between 2016 and 2024. Data collected included demographics, clinical presentation, radiological findings, operative details, adjuvant radiotherapy, endocrine outcomes, and metabolic complications.

Radiological evaluations included MRI brain in all patients, supplemented with CT scans when clinically indicated. Hormonal evaluation was performed preoperatively and at follow-up. Data were analysed descriptively.

Results

Demographics and clinical presentation

Of the 10 patients, 6 were boys and 4 were girls. The youngest was 3 years and the oldest 18 years.

Presenting symptoms included- Headache: 10 patients, Vomiting: 4 patients, Visual impairment: 2 patients, Seizures: 1 patient, Motor weakness (left upper and lower limb): 1 patient, Irregular menstrual cycles: 1 patient, Absent secondary sexual characteristics: 1 patient as in figure 1

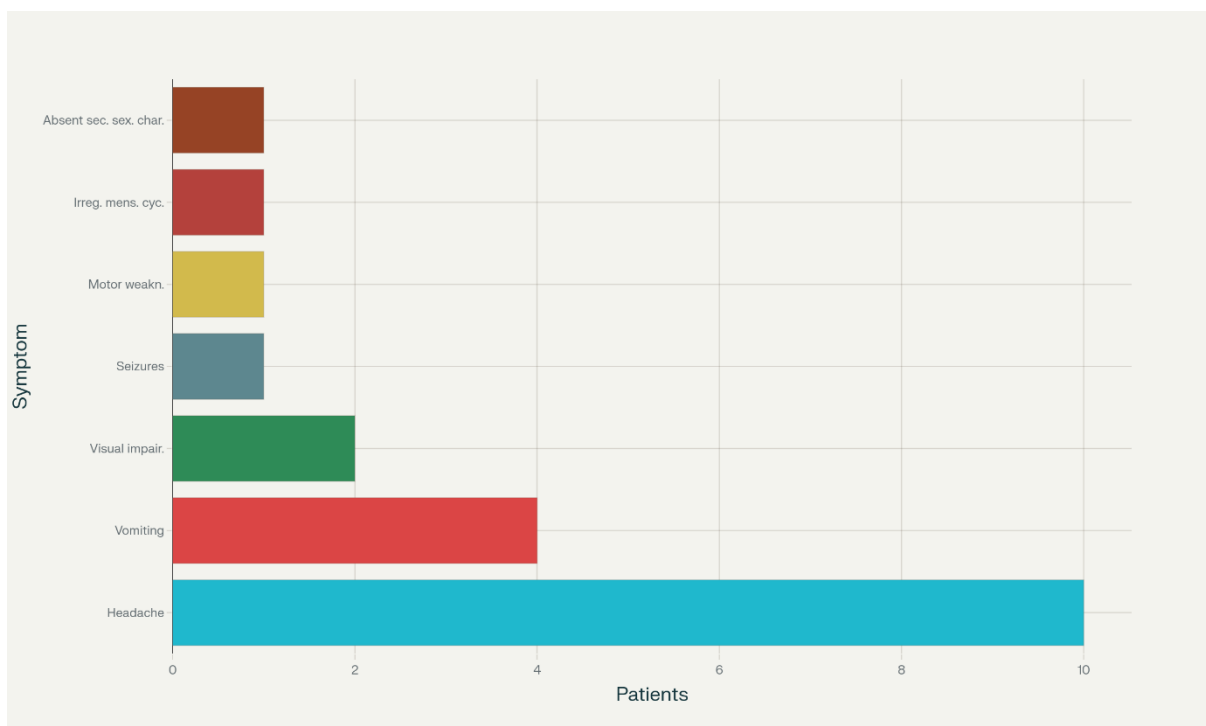


Figure 1: Presenting symptoms

On clinical examination, patient heights ranged from 96 cm to 158 cm, corresponding to growth below the 3rd centile in two patients, below the 25th centile in three patients, and below the 50th centile in five patients for their age and sex. Body weight varied from 14 kg to 82 kg, with BMI values between 15.2 kg/m² and 33.8 kg/m²; four patients were obese, four overweight, and only two within the normal range, indicating a high prevalence of excess

adiposity. Sexual maturity ratings revealed delayed or arrested pubertal development: in males, Tanner stages ranged from P1G1 to a maximum of P3G3, while in females stages ranged from B1P1 to B4P4, with no patient attaining full maturity. These findings are consistent with hypothalamic–pituitary axis dysfunction and growth impairment frequently associated with paediatric craniopharyngioma.

Table 1: Anthropometry, Sexual Maturity Rating, and Height SDS at Diagnosis (with specified SDS distribution)

Case	Age (yrs)	Sex	Height (cm)	Height SDS	Height Centile Category	Weight (kg)	BMI (kg/m ²)	BMI Category	SMR (Tanner stage)
1	3	M	96	-3.0	< 3rd centile	14	15.2	Normal	P1 G1
2	6	F	113	-3.0	< 3rd centile	23	18.0	Normal	B1 P1
3	8	M	120	-2.0	< 25th centile	36	25.0	Overweight	P1 G1
4	9	M	125	-2.0	< 25th centile	39	25.0	Overweight	P1 G1
5	10	F	130	-2.0	< 25th centile	48	28.4	Obese	B1 P1
6	11	M	135	-1.0	< 50th centile	42	23.0	Overweight	P2 G2
7	13	M	150	-1.0	< 50th centile	76	33.8	Obese	P3 G3
8	14	F	155	-1.0	< 50th centile	80	33.3	Obese	B1 P1
9	15	M	160	-1.0	< 50th centile	78	30.5	Overweight	P3 G3
10	18	F	158	-1.0	< 50th centile	82	32.9	Obese	B4 P4

Laboratory investigations are illustrated in table 2

Table 2: Baseline Hormonal Profile at Diagnosis

Case	8AM Serum Cortisol (µg/dL)	TSH (mIU/L)	Free T4 (pmol/L)	IGF-1 (ng/mL)	LH (mIU/ml)	FSH (mIU/ml)	Estradiol (pg/mL) / Testosterone (ng/dL)	Prolactin (ng/mL)
1	7.6	0.04	7.5	12	0.1	0.2	5 / —	28
2	6.5	0.12	8.0	14	0.2	0.3	8 / —	24
3	5.3	6.8	7.8	22	0.3	0.6	— / 20	18
4	6.0	5.9	9.2	24	0.4	0.8	— / 25	32
5	3.6	3.2	10.0	26	0.2	0.4	9 / —	15
6	4.8	0.08	8.3	32	0.5	0.7	— / 15	22
7	4.3	0.05	7.0	38	0.4	0.6	— / 120	26
8	7.6	0.09	8.1	38	1.2	2.0	14.2 / —	20
9	6.2	0.15	8.4	40	1.5	3.2	— / 250	17
10	4.7	0.07	7.2	42	1.8	4.2	120 / —	14

Pre-operative interventions- Ventriculoperitoneal (VP) shunt: 4, Endoscopic cyst aspiration: 1, Ommaya reservoir insertion: 2

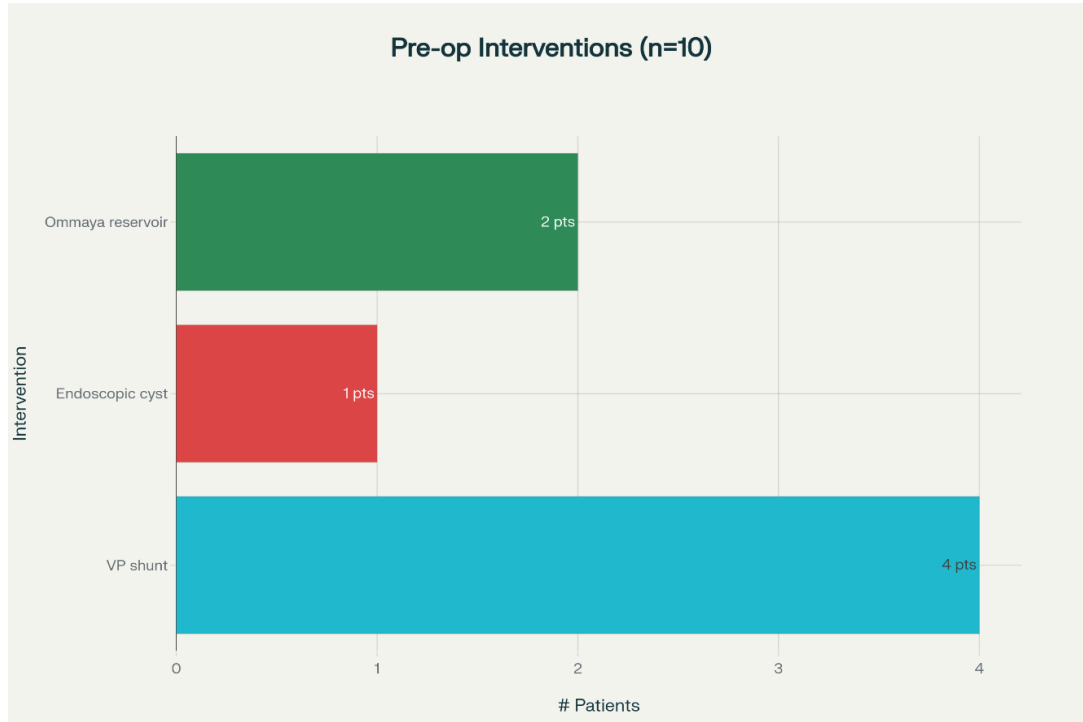


Figure 2: Pre-operative interventions

Imaging findings in figure-3 and 4, Post Ommaya reservoir placement CT-skull in figure-5 and Histopathology image of biopsy specimen in figure-6

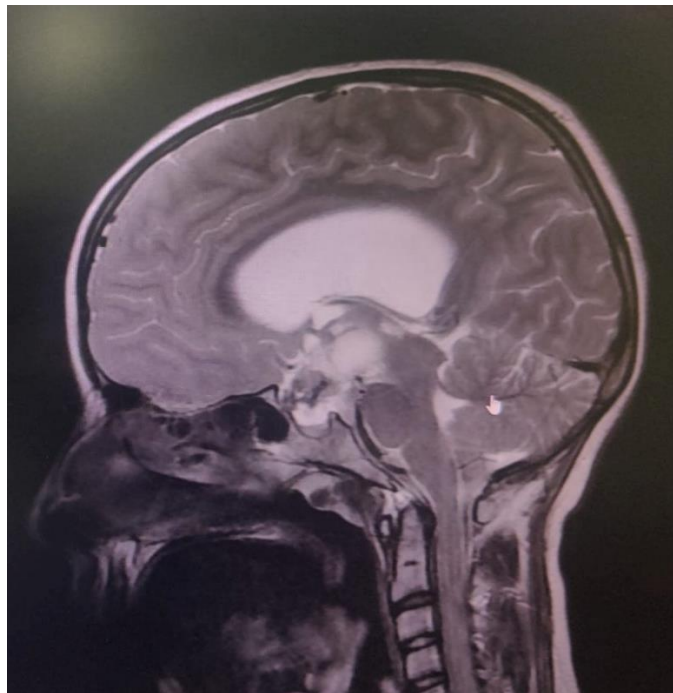


Figure 3 : MRI brain and pituitary -sagittal view of case 8 showing sellar and suprasellar lesion suggestive of Craniopharyngioma

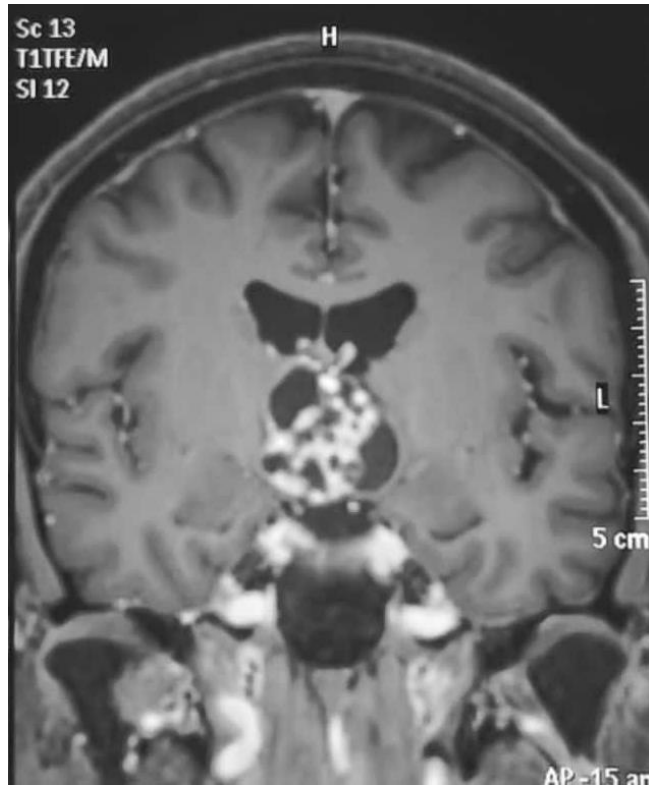


Figure 4 : MRI brain and pituitary – coronal view of case 10 showing sellar and suprasellar mass with cystic areas and calcifications suggestive of craniopharyngioma



Figure 5: CT skull lateral view- showing Ommaya reservoir

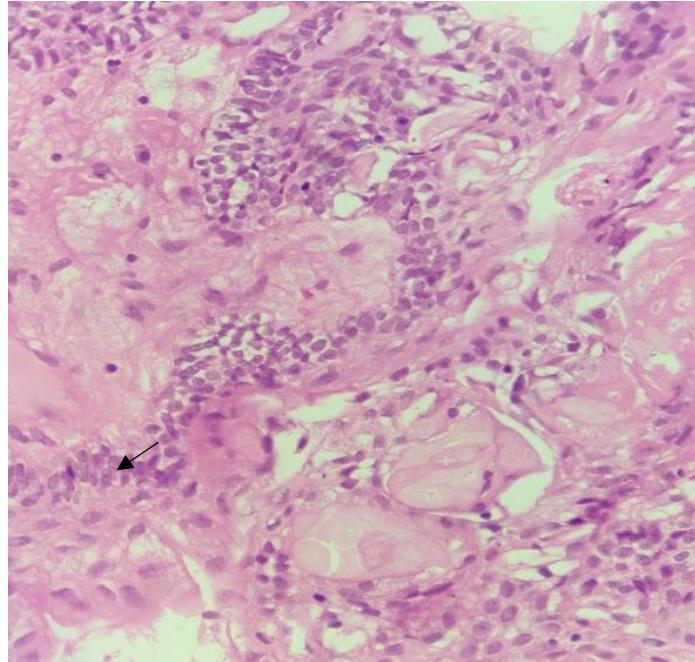


Figure 6: Histopathology showing wet keratin and cystic degeneration(arrow)- features suggestive of Adamantinomatous craniopharyngioma

Definitive treatment- All patients underwent Surgery, followed by: Conventional radiotherapy: 8, Stereotactic radiotherapy: 1, Proton beam therapy: 1(as in figure 3)

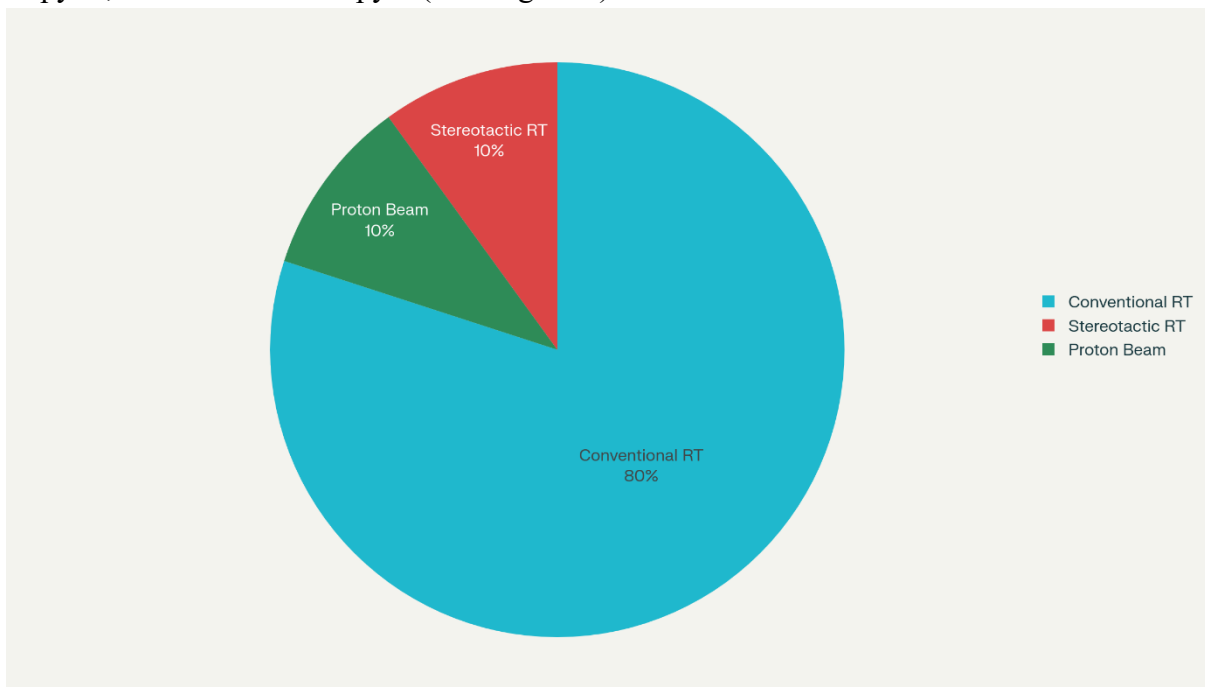


Figure 7: Type of Radiation Therapy

Hormone replacement therapy at 6 weeks post-surgery - Growth hormone:2, Levothyroxine:7, Glucocorticoid:8, Desmopressin:4, Sex steroid:3 (as in figure 4)

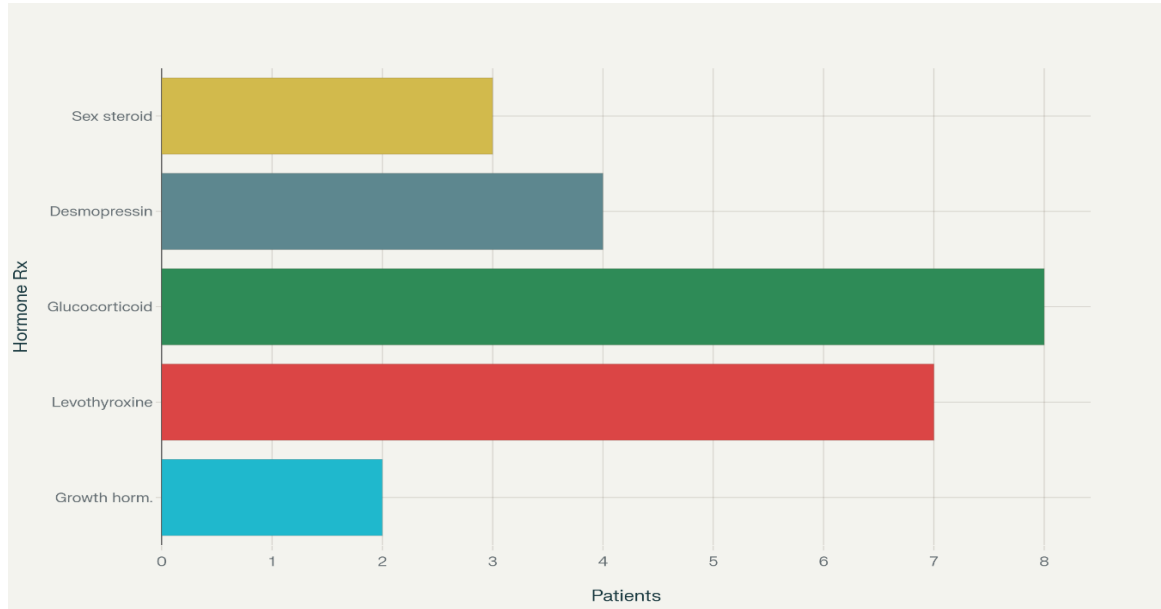


Figure 8: Hormone replacement therapy at 6 weeks post-surgery

Metabolic complications at follow-up

Out of the 10 patients-Obese: 4, Overweight: 4, Normal weight: 2, Impaired glucose tolerance: 1, Diabetes mellitus: 1, Hypertension: 1, Dyslipidaemia: 1

Metabolic Outcome	Number of Cases
Obese	4
Overweight	4
Normal Weight	2
Impaired Glucose Tolerance	1
Diabetes Mellitus	1
Hypertension	1
Dyslipidaemia	1

Survival outcomes

Median follow-up duration was 8 years. Seven patients are alive and progression-free; three patients died, two of them due to recurrence.

Discussion

Craniopharyngiomas, despite their benign histological nature, pose significant therapeutic challenges due to their location in the sellar/parasellar region and their propensity for local invasion and recurrence⁽¹⁾. Our series aligns with existing literature demonstrating that the majority of paediatric patients present with symptoms of raised intracranial pressure such as headache and vomiting, often necessitating cerebrospinal fluid diversion prior to definitive surgical management⁽²⁾. The management paradigm increasingly favours subtotal resection followed by adjuvant radiotherapy—

including conventional, stereotactic, and proton beam modalities—to balance effective tumour control with minimizing treatment-related morbidity⁽³⁾.

Endocrine dysfunction is a hallmark of craniopharyngioma and its treatments, with pituitary hormone deficiencies present in nearly all patients⁽³⁾. In our cohort, 80% required glucocorticoid or thyroid hormone replacement, reflecting common secondary adrenal insufficiency and central hypothyroidism⁽⁶⁾. Hypothalamic injury contributes further to metabolic complications such as hypothalamic obesity and metabolic syndrome, which substantially impact

morbidity and mortality^(4,5). Our observed high rates of obesity and overweight status are consistent with prior studies noting that up to 52–75% of paediatric patients develop hypothalamic obesity after treatment, underscoring the necessity of vigilant metabolic monitoring and management⁽⁸⁾.

Proton Beam Therapy for craniopharyngioma, especially in children and adolescents, has shown impressive local control and acceptable acute and late toxicities which included vascular and visual toxicities, hypothalamic obesity, endocrinopathy, and panhypopituitarism⁽¹²⁾.

Craniopharyngiomas are classified into adamantinomatous craniopharyngiomas (ACPs) and papillary craniopharyngiomas (PCPs) on the basis of their histological characteristics. Among them, ACP is more common, accounting for approximately 90% of all craniopharyngioma cases⁽¹¹⁾.

Survival rates for paediatric craniopharyngiomas are excellent in modern cohorts, with 5-year overall survival commonly reported between 80% and 95%⁽⁹⁾. However, the quality of life for survivors is often hindered by persistent endocrine, neurological, metabolic, and psychosocial sequelae⁽¹⁰⁾. These long-term complications necessitate multidisciplinary survivorship care incorporating endocrinology, neurosurgery, oncology, rehabilitation, and psychological support to optimize outcomes⁽⁷⁾.

Conclusion

Paediatric craniopharyngiomas have excellent long-term survival with modern multimodal treatment. However, high rates of panhypopituitarism and metabolic syndrome necessitate lifelong follow-up. Early intervention for cardiovascular risk factors and optimal hormone replacement can improve outcomes and quality of life.

List of Abbreviations

CSF – Cerebrospinal fluid

MRI – Magnetic resonance imaging

VP shunt – Ventriculoperitoneal shunt

ACPs - Adamantinomatous Craniopharyngiomas and

PCPs - Papillary Craniopharyngiomas

Declarations

Funding: None.

Conflicts of Interest: The authors declare no conflicts of interest.

Consent: Written informed consent for publication of anonymised data and images was obtained from guardians.

References

1. Steinbok P. Craniopharyngioma in children: long-term outcomes. *Childs Nerv Syst.* 2015;31(4):571-576. doi:10.1007/s00381-015-2643-x.
2. Bunin GR, Surawicz TS, Witman PA, Preston-Martin S, Davis F, Bruner JM. The descriptive epidemiology of craniopharyngioma. *J Neurosurg.* 1998;89(4):547-551. doi:10.3171/jns.1998.89.4.0547.
3. Müller HL. Craniopharyngioma. *Endocr Rev.* 2014;35(3):513-543. doi:10.1210/er.2013-1115.
4. Roth CL, Müller HL. Childhood craniopharyngioma and hypothalamic obesity: models for hypothalamic involvement in energy balance control. *Front Endocrinol (Lausanne).* 2010;1:63. doi:10.3389/fendo.2010.00063.
5. Mettal S, Bison B, Calaminus G, Warmuth-Metz M, Pietsch T, Kortmann R, et al. Metabolic syndrome and cardiovascular disease in craniopharyngioma patients. *J Clin Endocrinol Metab.* 2013;98(6):E1042-E1050. doi:10.1210/jc.2013-1179.
6. Children's Cancer and Leukaemia Group (CCLG). Craniopharyngioma: Guideline for the management of children and young people (CYP) aged <19 years UK: CCLG; 2021. Available at <https://www.cclg.org.uk/guidelines>
7. National Cancer Institute. Childhood craniopharyngioma treatment (PDQ®)– Health professional version. *PDQ Cancer Information Summaries.* Bethesda (MD):

- NCI; 2008. Available from: <https://www.cancer.gov/types/brain/hp/child-craniopharyngioma-treatment>.
8. Holmer H, Pozarek G, Wirfält E, Stibrant-Sunnerhagen K, Popovic V, Ekman B, et al. Hypothalamic obesity in adults with childhood onset craniopharyngioma and hypothalamic damage. *J Clin Endocrinol Metab.* 2009;94(9):3779-3786. doi:10.1210/jc.2009-0648.
 9. Tamburrini G, Caldarelli M, Massimi L, Santini P, Di Rocco C. Intracranial pressure monitoring in children with single suture and complex craniosynostosis: a review. *Childs Nerv Syst.* 2005;21(10):913-921. doi:10.1007/s00381-004-1117-x
 10. Del Baldo G, Vennarini S, Cacchione A, Amelio D, De Ioris MA, Fabozzi F, Colafati GS, Mastronuzzi A, Carai A. Multidisciplinary Management of Craniopharyngiomas in Children: A Single Center Experience. *Diagnostics.* 2022; 12(11):2745. <https://doi.org/10.3390/diagnostics12112745>
 11. An W, Li S, An Y, Lin Z. Molecular subtypes of adamantinomatous craniopharyngiomas. *Neuro Oncol.* 2025;27(5):1180-1192. doi:10.1093/neuonc/noaf030
 12. Li, Z., Li, Q., Tian, H. *et al.* Proton beam therapy for craniopharyngioma: a systematic review and meta-analysis. *Radiat Oncol* 19, 161 (2024). <https://doi.org/10.1186/s13014-024-02556-w>