Craniopharyngiomas - 20-year-period evaluation study

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Craniopharyngioma

"A histologically benign, partly cystic epithelial tumour of the sellar region presumably derived from embryonic remnants of the Rathke pouch epithelium, with two clinicopathological variants (adamantinomatous and papillary) that have distinct phenotypes and characteristic mutations."

-WHO Classification of Tumours of the Central Nervous System (2016)-

Craniopharyngioma

- WHO grade I.
- Constitute 1.2-4.6% of all intracranial tumours.
- The most common non-neuroepithelial intracerebral neoplasm in children (5-11% of intracranial pediatric tumours).
- Adamantinomatous subtype has a bimodal age distribution, with incidence peaks in children aged 5-15 years and adults aged 45-60 years.
- Papillary subtype occurs almost exclusively in adults, at a mean patient age of 40-55 years, without sex predilection.

Craniopharyngioma

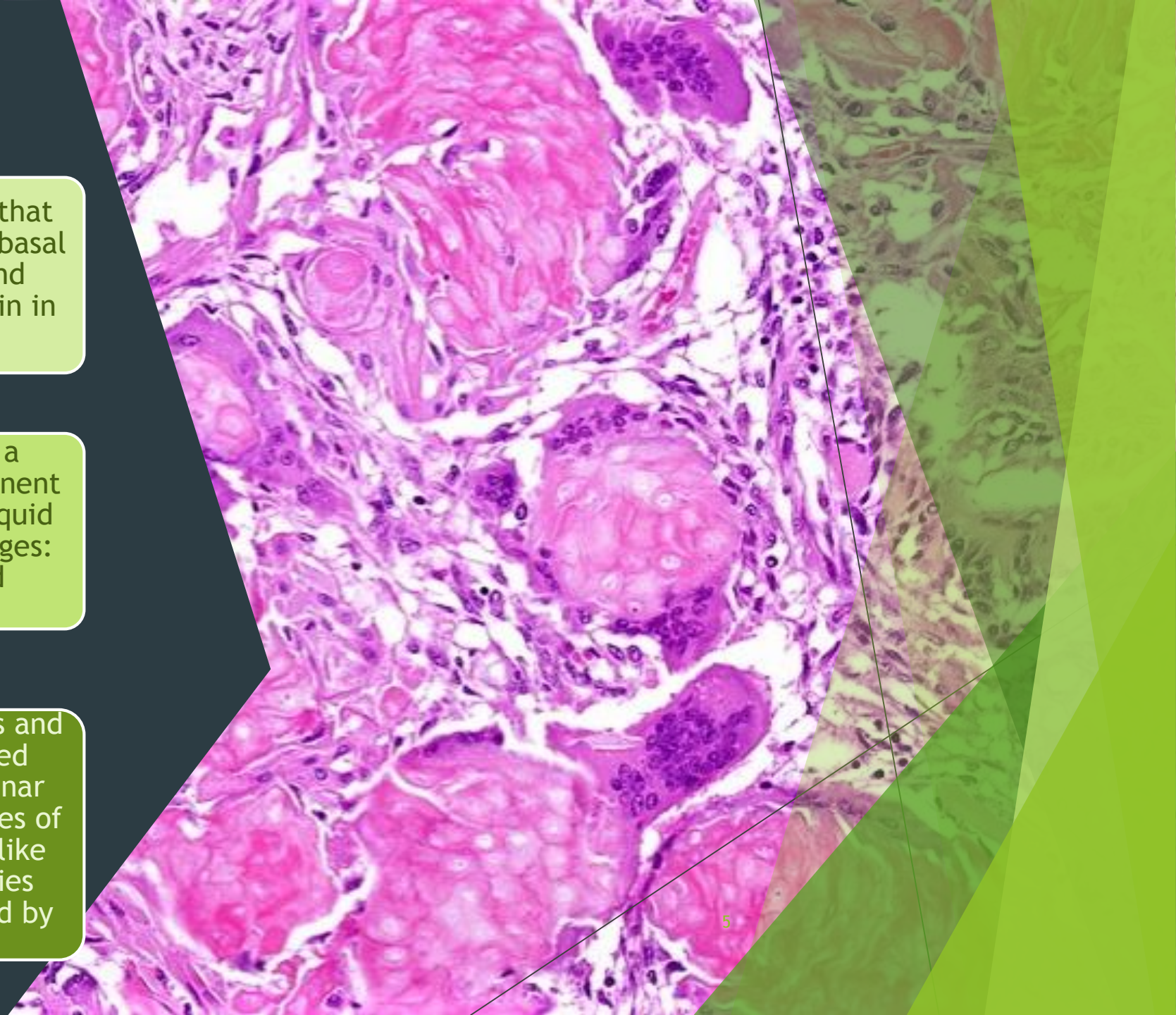
- The most common site for both subtypes is the suprasellar cistern.
- Unusual locations: sphenoid sinus and cerebellopontine angle.
- Papillary variant is also found in the third ventricle.
- Clinical features: visual deficits (adults); endocrine deficiencies (children): GH, LH/FSH, ACTH and TSH; diabetes insipidus; cognitive impairment and personality changes; hypothalamic dysfunction (obesity and hyperphagia).
- Frequent signs of increased intracranial pressure (cases with compression or invasion of the third ventricle).

Adamantinomatous craniopharyngioma

Characterized by a distinctive epithelium that forms stellate reticulum, wet keratin, and basal palisades, showing CTNNB1 mutations and aberrant nuclear expression of beta-catenin in 95% of cases.

Macroscopy: Lobulated solid mass with a spongy consistency - variable cystic component which may contain dark greenish-brown liquid resembling machinery oil. Secondary changes: fibrosis, calcifications, ossification and presence of cholesterol-rich deposits.

Microscopy: Cords, lobules, nodular whorls and irregular trabeculae of well-differentiated epithelium, bordered by palisading columnar epithelium - stellate reticulum. Pale nodules of wet keratin constituting anucleate ghost-like remnants of squamous cells. Cystic cavities containing cell debris and fibrosis are lined by flattened epithelium.

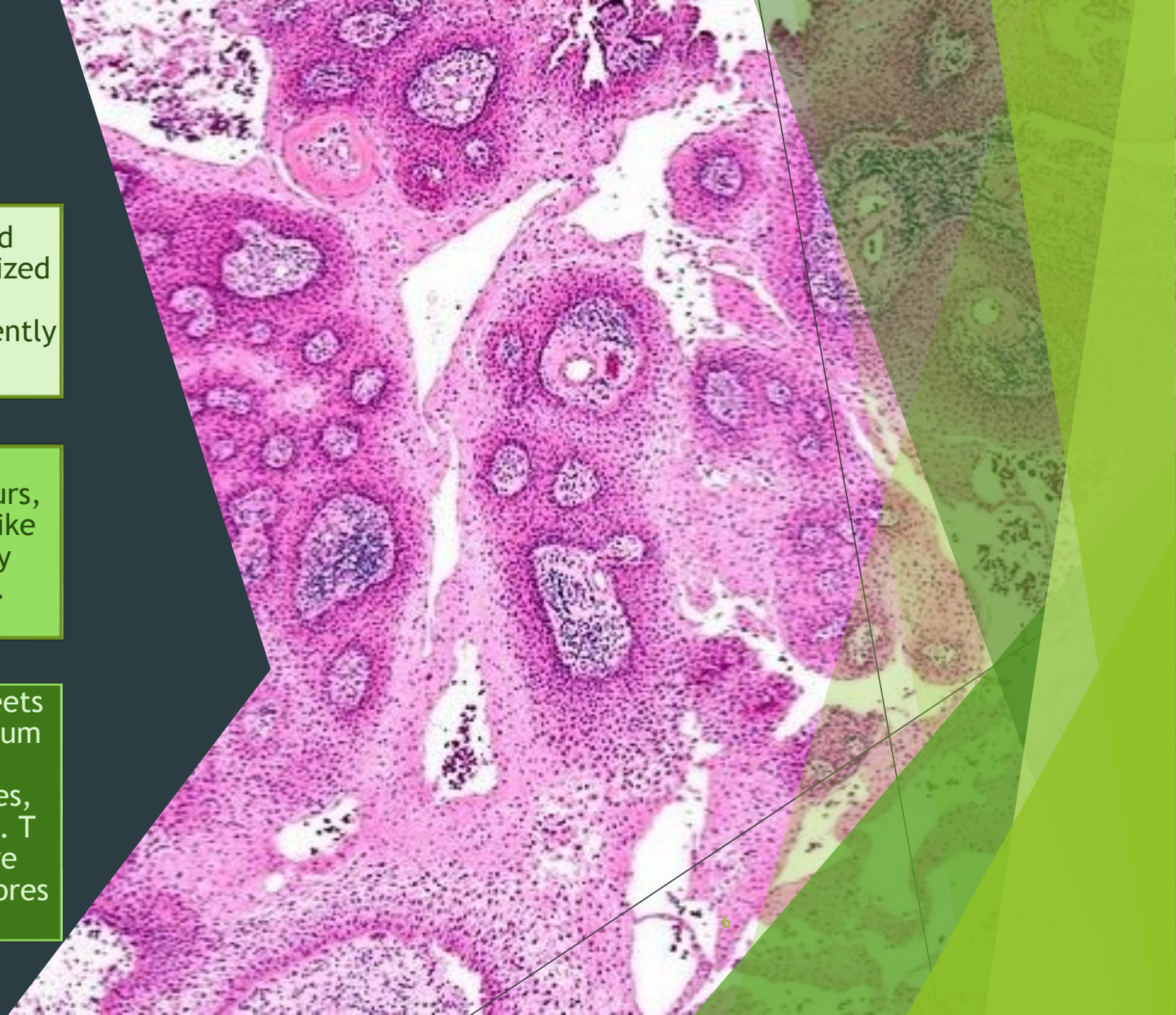


Papillary craniopharyngioma

Papillary, mostly supratentorial or third ventricular craniopharyngioma characterized by fibrovascular cores lined by non-keratinizing squamous epithelium. Frequently show *BRAFV*600E mutations.

Macroscopy: Solid or rarely cystic tumours, without cholesterol-rich machinery oil-like fluid or calcifications. The surface may appear corrugated or cauliflower-like.

Microscopy: Compact, monomorphic sheets of well-differentiated squamous epithelium without surface keratinization. Lacks calcifications, picket fence-like palisades, whorl-like cell nodules, and wet keratin. T cells, macrophages, and neutrophils are scattered throughout the fibrovascular cores and tumour epithelium.



Craniopharyngiomas: 20-year-period evaluation study

- Retrospective evaluation statistical study.
- Evaluation period: from 1998 to 2018 year.
- Patients were operated at the University Neurosurgery Clinic, Skopje, Macedonia.
- 40 craniopharyngioma cases (0.81% of the total 4929 diagnosed benign and malignant tumors) diagnosed on paraffin section slides, routinely stained with H&E at the Institute of Pathology, Medical Faculty - UKIM, Skopje, Macedonia.
- Statistica for Windows 7.

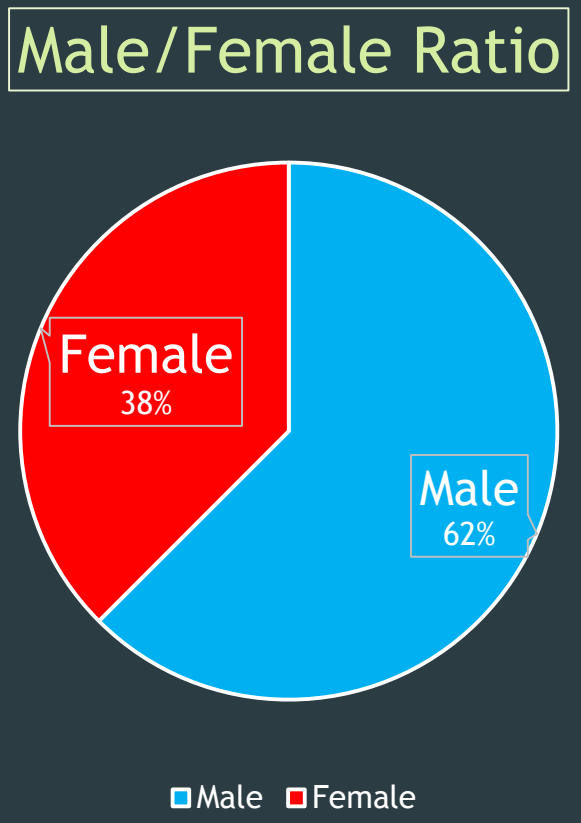
Craniopharyngiomas: 20-year-period evaluation study

Number of cases diagnosed annually/highest rate diagnosed in the 20 year period			
Year	**Number of cases**	**%**	**Incidence rate per 100,000 inhabitants**
1998	3	7.5	0,15
1999	5	12.5	0,25
2000	1	2.5	0,05
2001	2	5.0	0,10
2002	3	7.5	0,15
2003	2	5.0	0,10
2004	2	5.0	0,10
2005	2	5.0	0,10
2006	2	5.0	0,10
2007	2	5.0	0,10
2008	2	5.0	0,10
2009	3	7.5	0,15
2010	1	2.5	0,05
2011	2	5.0	0,10
2012	1	2.5	0,05
2013	2	5.0	0,10
2014	2	5.0	0,10
2015	1	2.5	0,05
2016	1	2.5	0,05
2017	1	2.5	0,05
Total	40	100%	Prevalence 0,10 /100 000 inhabitants

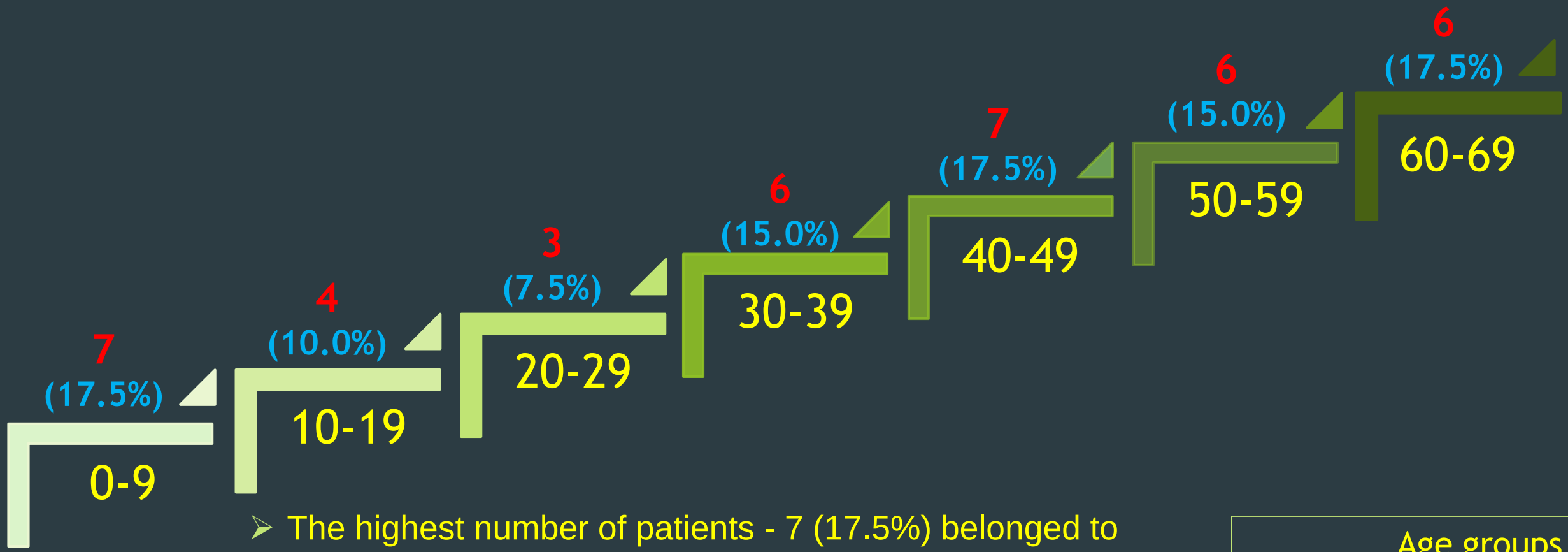
- The prevalence rate of diagnosed craniopharyngiomas is 0.10 per 100 000 inhabitants.
- Most, 5 (12.5%) cases were registered in 1999, with an incidence rate of 0.25 per 100 000 inhabitants in that year.

Craniopharyngiomas: 20-year-period evaluation study

- 25 (62.5%) males; 15 (37.5%) females.
- Average age: 36.4 ± 20.7 years.
- Youngest patient: 3 years.
- Oldest patient: 68 years.
- Average age: men 37.9 ± 17.8 years, women 34.0 ± 25.4 years.
- There is no significant difference in age between men and women with diagnosed craniopharyngioma (Mann-Whitney U test: Z = 0.489 p = 0.6246).



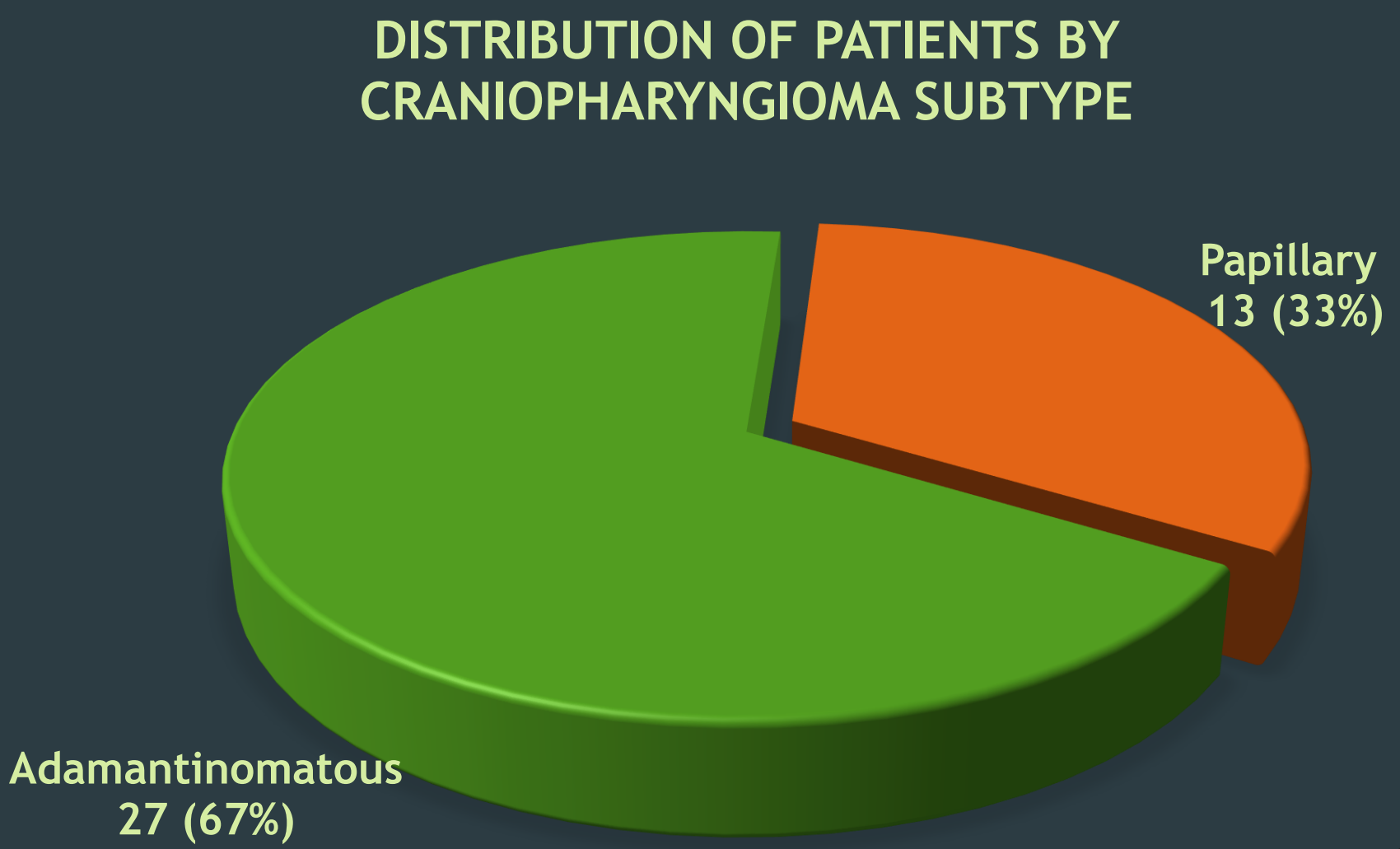
Distribution of patients by age groups



- The highest number of patients - 7 (17.5%) belonged to the age groups from 0 to 9, 40 to 49, and 60 to 69 years, and the lowest number - 3 (7.5%) were at the age of 20 up to 29 years.

Age groups
Number of patients
(%)

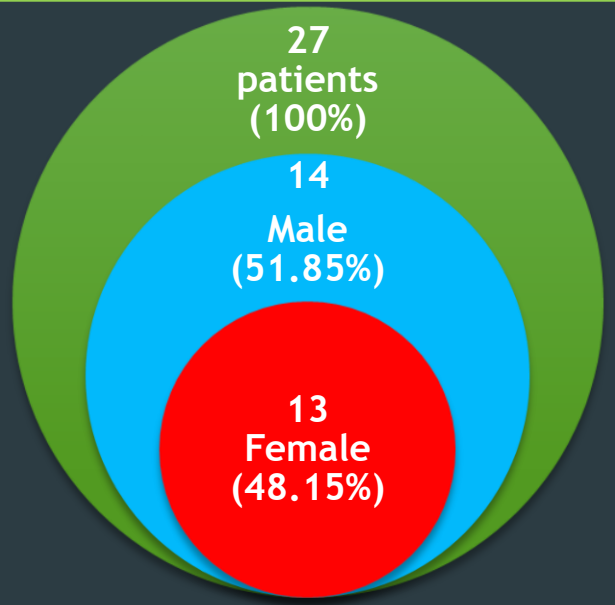
Craniopharyngiomas: 20-year-period evaluation study



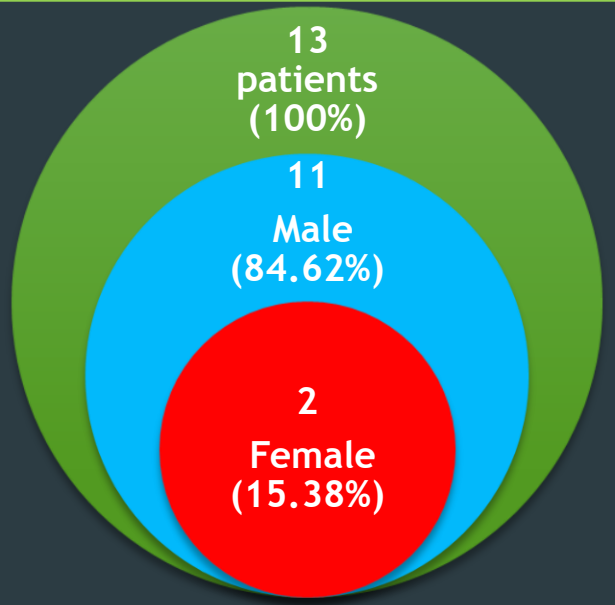
Craniopharyngiomas: 20-year-period evaluation study

Distribution of patients according to the craniopharyngioma subtype and gender

Adamantinomatous craniopharyngioma

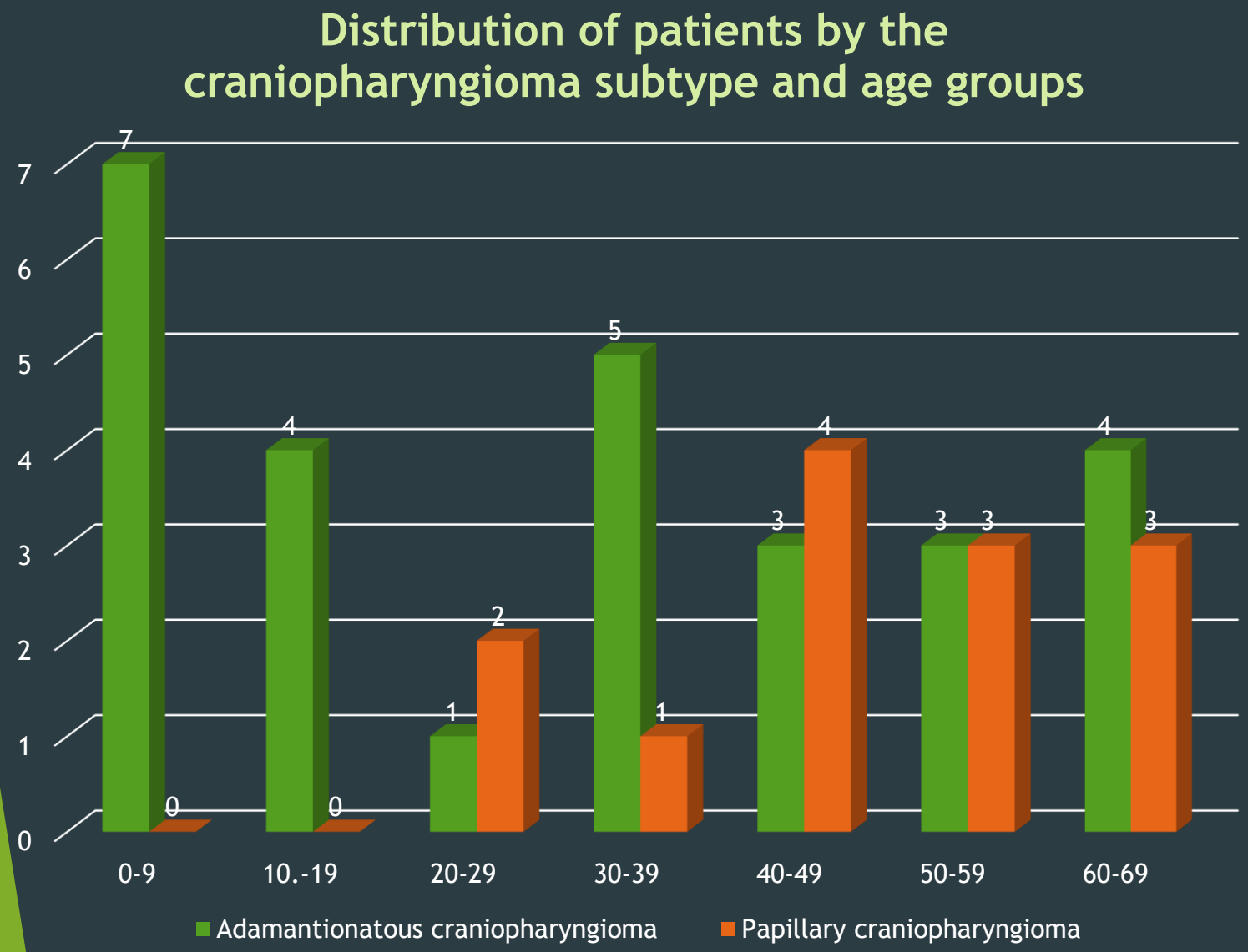


Papillary craniopharyngioma



- Adamantinomatous craniopharyngioma was diagnosed in 14 (51.85%) male and 13 (48.15%) female patients.
- Papillary craniopharyngioma was diagnosed in 11 (84.62%) male and in 2 (15.38%) female patients.
- There is an association/correlation between the patient's gender and the craniopharyngioma subtype (Fisher exact test: $p = 0.0458$). Papillary craniopharyngiomas are significantly more common in men.

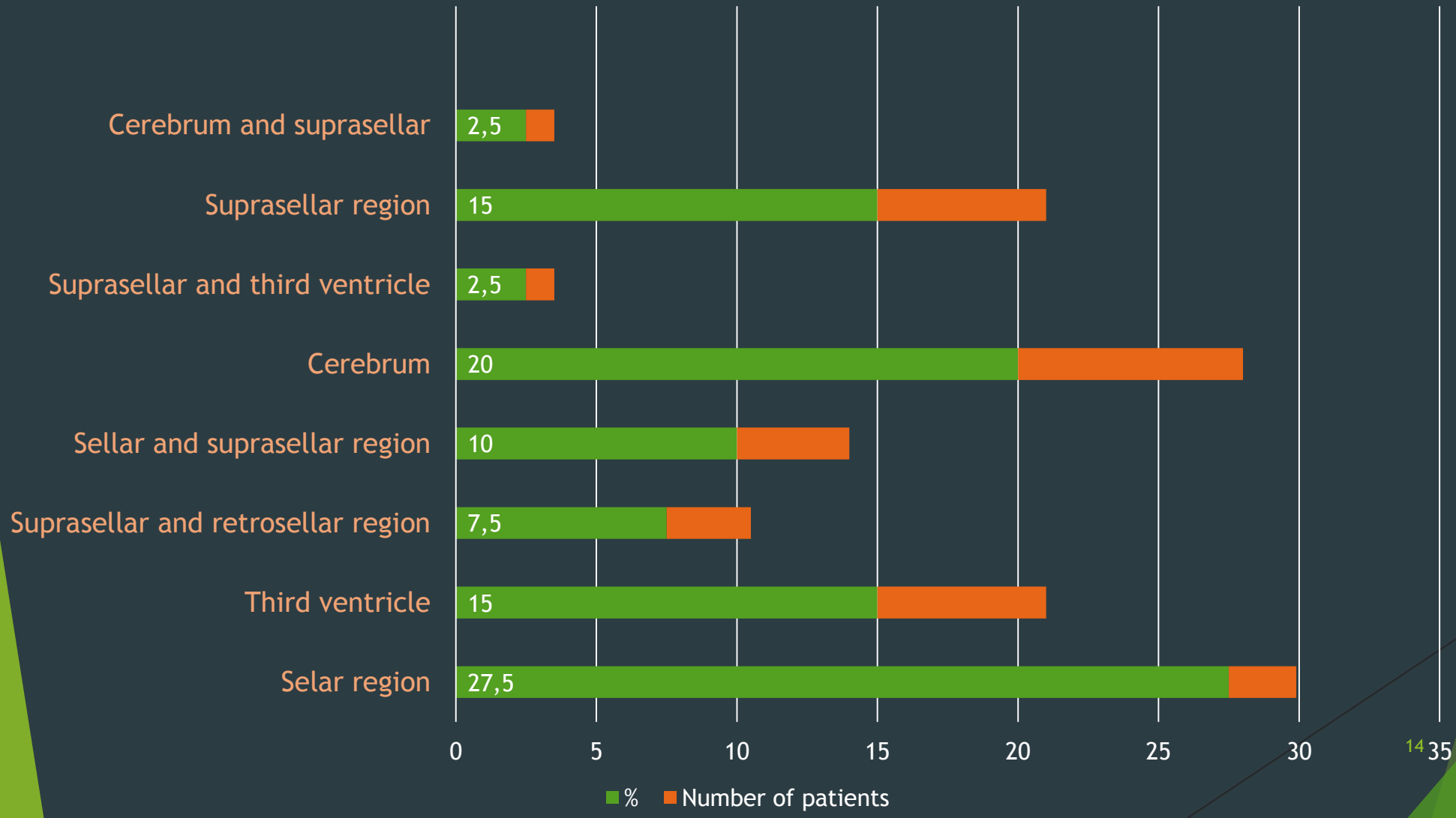
Craniopharyngiomas: 20-year-period evaluation study



- There is a statistically significant difference between age groups in relation to the craniopharyngioma subtypes (Kruskal-Wallis ANOVA: H = 14.86 p = 0.0274).
- Adamantinomatous craniopharyngioma is significantly more pronounced in 0 to 19 age groups.

Craniopharyngiomas: 20-year-period evaluation study

Distribution of patients by tumor localization



Recurrence/Relapse

- Of the total of 40 patients with craniopharyngioma diagnosed during the evaluation period, 9 (22.5%) have experienced relapse of the tumor for a period of 1 to 3 years.
- The earliest relapse was in the same year of the primary diagnosis, and at the latest was after 3 years.
- In 6 patients of this group, there was 1 relapse, in 1 patient there were 2 relapses, and 2 of the patients experienced 3 relapses.

Conclusions

- Craniopharyngiomas are representing 0.81% of the total 4929 diagnosed benign and malignant tumors in the 20-year period in the Republic of Macedonia with a prevalence rate of 0.10 per 100 000 inhabitants.
- There is no significant difference in age between men and women with diagnosed craniopharyngioma.
- The highest number of patients (17.5%) belonged to the age groups 0-9, 40-49, and 60-69 years, and the lowest number (7.5%) belonged to the 20-29 age group.
- There is an association/correlation between the patient's gender and the type of craniopharyngioma. Papillary craniopharyngiomas are significantly more common in men.
- Adamantinomatous craniopharyngioma is significantly more pronounced in young people - aged 0 to 19 years.

Thank you for your attention