List the tumours that may arise in CPA:

1. Vestibular schwannoma: 75-90%
2. Meningioma: 5-10%
3. Epidermoid 5%
4. Cholesteatoma: 5%
5. Other schwannomas 2-5%: trigeminal is the most common (0.3% of intracranial tumours), followed by facial, vagus, glossopharyngeal, chorda tympani, tympanic branch of IX (Jacobson) and auricular of X (Arnold).
6. Metastases:1-2%
7. Glomus jugulare tumour
8. Tumours extending from cerebellum or 4th ventricle or brain stem (exophytic glioma, ependymoma)
9. Others: Dermoid, lipoma, chondroma, chondrosarcoma, chordoma

List other sites at which cranial schwannomas can occur and compare and contrast with CPA schwannomas:

1. **Trigeminal schwannoma**: 0.3% the second most common intracranial schwannoma after vestibular schwannoma. They can arise from the nerve root 2.2 mm distal to brain stem (start of Schwann cells), from the ganglion (most common) and from the divisions. 30% in posterior fossa , 50% middle fossa and 20% dumbbell

    - Clinical presentations: hypoesthesia in the distribution of V1-3, trigeminal neuralgia. Posterior fossa lesions can present with CPA mass signs (hearing loss, ataxia, brain stem compression), middle fossa lesions may present with cavernous sinus syndrome.
    - Diagnosis: radiological appearance is similar to vestibular schwannoma. Dumbbell lesions can cause erosion of the petrous apex with posterior and middle fossa components.
    - Gasserian ganglion lesions can be approached through subtemporal intradural approach, pure posterior fossa lesions can be approached through suboccipital craniotomy and dumbbell lesions require combined petrosal approaches with division of the tentorium.

2. **Facial schwannomas**: 2% of all intracranial schwannomas (third most common): The tumour can occur in any of the 5 segments of the facial nerve (cisternal, canalicular, labyrinthine (under the cochlea and vestibule and the shortest 3-4 mm), tympanic and mastoid) most commonly near the geniculate ganglion. The nerve acquires Schwann cells 2mm distal to the origin from brain stem.

- The most common presentation is hearing loss (sensorineural if the lesion from the first 2 segments and conductive if the lesion in the last 2 segments). Other symptoms include facial weakness, hemifacial spasm, and otalgia. Large lesions can cause cerebellar and brain stem signs.

- Diagnosis: enlargement of one of more of the segments of facial (fallopian canal) with enhancing mass on CT and MRI.

- The choice of the surgical approach depends on the location of the tumour and the status of hearing. If hearing is lost and the lesion is proximal to geniculate ganglion-translabyrinthine approach with nerve repair or grafting. If hearing is preserved –subtemporal or suboccipital approach depending on the location of the lesion.. Tumours in the last 2 segments can be removed through simple mastoidectomy.

3. **Rare locations**: jugular foramen (glossopharyngeal, accessory and vagus), Meckel's cave 3-d, 4^{th} and 6^{th} nerve in the cavernous sinus (extremely rare) and orbit.

Acoustic neuroma (vestibular schwannoma):

- Benign slow growing tumours of the Schwann cell at the junction of the central and peripheral myelin (8-12 mm from brain stem) usually start in the IAM.

- 6-8% of intracranial tumours, 60-80% of C-P angle tumours. 5% are bilateral. Peak age-fourth and fifth decade.5% of patients have NF2. In patients with NF2 the majority present before 30. M: F 43%:57%.

- The incidence is higher in autopsy studies (2.5%) which may indicate that some lesions remain asymptomatic. 50-60% arise from superior vestibular nerve, 40-50% inferior vestibular nerve and 10% from cochlear nerve.

- The growth rate is variable with the majority of tumours having slow growth< 2mm/per year, 12% remain static and 6% even decrease in size. Faster growth in patients with NF2. Enlargement of the tumour can be due to proliferation, haemorrhage and cystic degeneration. The growth rate of bilateral tumours is faster than for unilateral ones.

- In the majority the growth pattern can be predicted after 2-3 years of follow up(not always)

- Pathology: encapsulated, firm, lobulated tumour that distorts rather than invades the brain, expands the IAM in 90%. Microscopically biphasic tumour with **Antoni A areas** composed of spindle cells with nuclear palisading (**Verocay bodies**) and **Antoni B areas** which are less cellular with **pleomorphic, vacuolated cells in eosinophilic background** and microcystic areas. Stain +ve for S-100 (not specific, meningiomas, melanomas). Tumours in NF2 tend to invade the nerves and to grow at faster rate. **Ancient changes such as pleomorphism and necrosis** are degenerative changes and are not indicators of malignancy. Malignant vestibular schwannomas are extremely rare with only handful of reported cases. **45% contain estrogen or progesterone receptors**, hence the increased growth during pregnancy.

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- Presentations:

1. Hearing loss mostly gradual (in 10-20% sudden in onset due to vascular occlusion).SD is affected more than the pure tone loss

2. Tinnitus : the presenting symptom in 3% and is present in 60-80% of patients

3. Disequilibrium and vertigo Gradual loss of the function of one vestibular nerve is not disabling (compensation by the contralateral site)

4. If the tumour is large it can cause **hydrocephalus, cerebellar signs (**ataxia and nystagmus), brain stem compression signs (long tract signs, optokinetic nystagmus and cranial nerve palsies) and V, VII (weakness and hemifacial spasm), and lower cranial nerves symptoms and signs. Rarely the tumour can be intralabyrinthine.

Acute neurological deterioration can be due to haemorrhage or cystic expansion.

- Investigations:

1. CT scan with bone windows shows: iso or hypodense lesion with homogenous enhancement, **expansion of IAM**. Look for the degree of **temporal bone airation** which gives idea about the location of jugular bulb, less sensitive than MRI in showings small lesions. Poorly pneumatised temporal bone may indicate high jugular bulb while well pneumatised temporal bone increases the risk of CSF leak.

2. MRI with contrast is the method of choice .It can detect **lesions 2-3 mm** large. Enhancing lesion (inverted cone appearance). False positive MRI has been reported due to arachnoiditis and adhesions. Tumours are

classified into intracanalicular, small< 1 cm, medium size 1-2.5 and large > 2.5 cm

3. Audiometry: **speech recognition threshold** (SRT) and **speech discrimination SD** (sensorineural hearing loss at high frequency ,SD is more affected)

4. Auditory Brain stem responses: 100% sensitivity, 80% specificity can be used as screening test with audiometry and caloric testing

5. Electronystagmography: demonstrates impaired vestibular function in 80% of patients

Nystagmus in vestibular schwannoma can be **horizontal** (vestibular) with the fast component away from the lesion, **optokinetic** (using rotating object)-gaze center compression in the pons

Discuss the association of vestibular schwannoma with other lesions including NF2 and the underlying genetic abnormalities:

1. 5% of patients with acoustic schwannoma have NF2 and bilateral acoustic schwannomas are the whole mark of NF2

2. Genetics: **Mutations in NF2 gene on the long arm of chromosome 22(**missense and splice site mutations**)** and loss of heterozygosity on chromosome 22 are found in both sporadic (44%) AS and in patients with NF2 (100%). In **NF2 there is germ line** transmission of the mutation with second hit mutation in the second allele later on in life, while in sporadic schwannomas; mutations develop in both gene alleles later in life. Nf2 gene is a tumour suppressor gene that codes for **Merlin** which acts as linker between cell membrane and cytoskeleton. The mechanisms of tumour suppression are not known

3. Mosaicism of NF2 (unilateral vestibular schwannoma and unilateral tumours" meningiomas, astrocytomas and so on".

4. Increased incidence of meningiomas in patients with AS even in the absence of NF2 (loss of heterozygosity of chromosome 220.

- Treatment options are :

1. **Conservative treatment** with follow up by neuroimaging (in patients **with small, incidental tumours**, particularly in **elderly with medical problems** and in patient **in whom the tumour affects the only one hearing ear**). This is supported by the following

A. The majority are slow growing tumours with 70% growing< 2mm per year, 12% no growth and 6% negative growth

B. The growth pattern can be predicted in the majority of cases after a period of follow up (2-3 years)

C. Big discrepancy between the incidences in clinical studies 9/1000000 and autopsy studies 2.5% which indicates that many tumours remain asymptomatic and have a benign course

D. Surgery carries risk of mortality and morbidity. Hearing preservation rate in best series is about 33%. With radiosurgery delayed hearing loss occurs in 25-75% of patients

2. **Surgical excision** through retrosigmoid, translabyrinthine and middle fossa approaches .(read Kaye for details)

A. Retrosigmoid approach has the following advantages: familiar to neurosurgeons, can be used to remove tumours of any size, can be used for hearing preserving operation. Disadvantages include the need for cerebellar retraction and the difficulty in visualising the fundus of IAM, the facial nerve is initially out of view (displaced anteriorly by the tumour)

B. Translabyrinthine approach: advantages include minimal cerebellar retraction; can be used for removing large tumours, early identification of VII nerve. Disadvantages: hearing loss and higher incidence of CSF leak and meningitis.

C. Middle fossa approach: advantages : hearing preservation, disadvantages: temporal lobe retraction (haemorrhage , epilepsy, dysphasia), suitable only for small tumours and tumours with CPA extension < 1cm, potentially higher incidence of facial nerve palsy because the facial nerve is in the upper part of the exposure and one has to work around it.

3. **Stereotactic radiosurgery**: is a treatment option for

A. lesions smaller than 3 cm,

B. Residual tumours after surgical resection

C. Tumours affecting the only hearing ear. NF2

D. Recurrent lesions.

The first published report of radiosurgery being used to treat acoustic neuromas was by Leksell in 1971. Since then, multiple retrospective studies have documented the high likelihood of tumour control with such treatment Overall rates of tumour control have been shown to be **92 to 100%** in the first several years after radiosurgery) and **98%** after 5 to 10 years of follow-up. The initially used dose was **15-25 grey** to the center and **10-15 greys** to the tumour periphery. This dose was associated with20% facial nerve paresis and 50% and 75% hearing loss at 1 and 2 years.

Strategies used to decrease the risk of complications:
1. **Decrease in the single-stage** radiosurgical dose for acoustic neuroma from >14 Gy to **<14 Gy has nearly eliminated radiation-induced facial nerve injury and has increased the rate of hearing preservation**. Contemporary rates of hearing preservation have been reported to be 71 to 73%

2. **Fractionated stereotactic radiotherapy**. Several preliminary and retrospective studies have reported improved rates of hearing preservation after standard fractionated treatment of acoustic neuroma, from **68 to 100**% at 18 to 23 months and 71 to **85% at 2 and 5 years**

Discuss the indications for and types of adjuvant therapy for CPA tumours and related pathological types:

1. External beam radiotherapy for residual and recurrent acoustic schwannoma: rarely used now a day. Some authors reported that postoperative radiotherapy reduced the risk of recurrence of AS from 46% to 6%. RT has a role in chordoma, chondrosarcoma, metastasis and lymphoma (se above).
2. Stereotactic radiosurgery can be used as a primary treatment or as an adjuvant treatment for residual or recurrent tumour.
3. Chemotherapy: has a role in the treatment of lymphoma and metastases. No role in the treatment of schwannomas.

Describe the House –Brackmann grading system for facial nerve weakness: (Kaye page 627): assess the function at rest and with motion (forehead, eye closure and mouth symmetry and weakness).

Difficult to differentiate grade 2 from 3 and 4 from 5

4. Grade 1-normal facial function in all areas.
5. Grade 2 –mild dysfunction: slight weakness noticeable at close inspection. Normal symmetry and tone at rest, normal eye closure with minimum effort. Slight mouth asymmetry.
6. Grade 3 moderate dysfunction: at rest –normal symmetry and tone. Motion: forehead-mild movement, eye-complete closure with effort and mouth slightly weak with max. effort.

7. Grade 4: moderate severe dysfunction: at rest-obvious weakness and asymmetry. With motion-Forehead-no movement, eye-incomplete closure, mouth-asymmetric with maximum effort
8. Grade 5: severe weakness- at rest: obvious asymmetry. On motion: forehead-no movement, eye : incomplete closure with minimal movement, mouth-slight movement
9. Grade6-total paralysis with no movement.

Discuss the management of a patient with facial paresis:

Postoperative facial nerve function depends on the size of the tumour (**H-B grade 1-2 in 70-90% of patients with small tumours and only in 30% of large tumours > 3cm** in most recent series. Other factors influencing facial nerve function are surgeon's experience and the use of continuous EMG facial nerve monitoring.

I. **Facial nerve is in continuity**: some return of function can be anticipated in 90% of cases. Until then the primary goal is the prevention of corneal ulcer(incomplete eye closure, hypoesthesia from 5th nerve injury and decreased lacrimation from SGPN dysfunction)

1. Methyl cellulose artificial tears
2. Lacri-Lube ointment
3. Eye shield (avoid patches-contact with cornea)
4. Lateral tarsorrhaphy(temporary)
5. If no improvement in 1 year consider permanent tarsorrhaphy or upper eyelid gold weights insertion with medial canthoplasty, palpebral spring implantation.
6. Delayed hypoglossal-facial anastomosis.

II. **Facial nerve divided** : (Kaye page 659 for details)

1. Primary nerve repair (end-end anastomosis). Advantages : the function of other nerve is not sacrificed, reported good results in 60%
2. Repair with interposition graft (sural, posterior auricular). If the nerve ends cant be brought together.
3. Hypoglossal-facial anastomosis(partial or complete
4. Cross facial anastomosis with interposition graft