# Cerebellopontine angle Choroid plexus papilloma various presentations: report of series and literature review

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# Abstract

Choroid plexus tumors (CPTs) are rare choroid plexus epithelium originated neoplasms. Among all, choroid plexus papilloma (CPP) is the most common type of CPT. Clinical features and signs at diagnosis of CPP in CPA are so extensive and also nonspecific. This work aimed to review the sign and symptoms of CPP tumors in CPA through all cases reported in the literature. In this study, Data regarding presenting symptoms of 60 adult patients (including 53 adult patients identified from the literature and our seven new patients that presented in this study) with CPP in CPA were reviewed. Symptoms related to 8th cranial nerve, lower motor, 7th, and 5th cranial nerves were the most prevalent symptoms respectively following ICP rising and cerebellar signs. The most common symptoms were related to the ICP elevation, cerebellar and auditory system involvement. The rarity of CPP tumors in CPA along with a variety of presenting symptoms makes the diagnosis of CPPs more challenging while considering this variance could be helpful in prompt diagnosis.


# Introduction

Choroid plexus tumors (CPTs) are rare intraventricular neuroectoderm neoplasms derived from the choroid plexus epithelium [24,32,36]. According to the 2007 WHO Classification of Tumors of the Central Nervous System, primary CPTs are classified as choroid plexus papilloma (CPP, WHO grade I,80%), atypical CPP (WHO grade II,15%) and choroid plexus carcinoma (CPC, WHO grade III, less than 5%) [8,18]. Choroid plexus papilloma (CPP, WHO grade I) represent 0.4- 0.6 % of all primary intracranial tumors and occur more frequently in children [5]. The average annual incidence of all CPTs is approximately 0.3 per 1 000 000 people [39]. These tumors predominantly arise in supratentorial locations such as lateral ventricles in children and the fourth ventricle in adults [27,29]. CPPs developing in the cerebellopontine angle (CPA) are uncommon and are almost always found in adults [25,31,33,38]. They represent about 9% of all CPPs and are almost exclusively found in adults [5,18,24]. Clinical features and signs at diagnosis of CPP in CPA include the ICP rising symptoms, gait impairment, cranial nerve palsies (V, VII, VIII), seizures, cerebellar signs, and psychomotor retardation [14,17].

CPP tumors are not among common tumors of CPA, although in patients who have a combination of unusual symptoms could be one of the most important differential diagnoses. This study aimed to review the signs and symptoms of CPP tumors in CPA through all adult cases reported in the literature to give a more unobstructed view of these tumor presentations.

# Method

# Patients Series

We retrospectively collected data of a cohort of consecutive patients operated on for primary choroid plexus papilloma in CPA between 2011 and 2018 in the Department of Neurosurgery at the Loghman

Hakim Hospital affiliated to Shahid Beheshti University of Medical Sciences. Only adult patients with primary tumors arising from the CPA were included. All tumors with secondary CPA involvement were excluded. The collected data included presenting clinical signs and symptoms, the surgical approach, and post-surgical follow up. We reviewed the medical records retrospectively. Due to the data being deidentified, no ethics committee approval was needed.

# Literature review

To perform an updated review on different symptoms and signs of choroid plexus papilloma of CPA, in adult patients, we carried an electronic literature search among articles published in English prior to March 2019. Articles were retrieved from PubMed, Scopus, EMBSCO, CINAHL, ISC and the Cochrane Library using keywords including “Choroid plexus papilloma”, “Cereberopontine angle”. The integrative review included 17 studies until March 2019 that are presented and discussed as follow.

## Results

This study included 7 adult patients with CPP in CPA who were treated surgically at our institutions between 2013 and 2017 (Table 1). There were 4 male and 3 female patients, and the mean age was 40.57 years (range 32–47 years). The average follow-up period after surgery was 16.2 months (range 12–24 months). Follow-up was completed in all cases because no evidence of recurrence was observed for 12 months after surgery.

In addition to all related articles, additional cases from their references were reviewed. A total of 17 articles were included in this review.

# Literature review

Sixty-eight cases were derived from the literature, either published as a case report or mentioned in a larger case series focusing on a different research question. Of these 68 cases, 15 were younger than 18 are excluded. Therefore, 60 cases (including our 7 cases) are included in this study.

# Demographics

The complete series (both our series and the cases identified in the literature) consisted of 37 females and 23 males. Mean age of cases was 40.78 (median: 40), and there was no significant difference between male (average: 40.7; median: 40) and female (average:40.8; median: 39) patients.

# Presenting Symptoms

To facilitate the review, the clinical symptoms of patients were divided into 7 categories including symptoms associated with increased intracranial pressure (ICP), cerebellar symptoms, auditory symptoms, symptoms related to the lower motor system, symptoms pertained to the nerve VII involvement, manifestations related to the nerve V involvement, and hemiparesis (Table 2). Patients with CPP in CPA had a wide range of clinical symptoms, most commonly associated with an increase in ICP (in almost all patients), symptoms related to cerebellar damage, auditory damage, nerve VII involvement, and other manifestations, each of which may be seen in other tumors of this area with a different pathology.

As shown in Figure 1, among the most important clinical symptoms mentioned in 60 patients, the highest prevalence of clinical manifestations of CPP in CPA is related to the increased ICP (51 cases, 85%), cerebellar involvement (32 cases, 53.33%), auditory system involvement (23 cases, 38.33%), nerve VII involvement (13 cases, 21.66%), lower motor (14 cases, 23.33%), nerve V involvement (9 cases, 15%), and hemiparesis (5 cases, 8.33%). Furthermore, among all symptoms which mentioned in all reports, symptoms related to 8th, lower motor, 7th, and 5th cranial nerves were the most prevalent symptoms respectively following ICP rising and cerebellar signs (Figure 2). This frequency is perfectly matched with the anatomical proximity to the Foramen Luschka.

# Discussion

Cerebellopontine angle (CPA) is the most common site for posterior fossa tumors, where generally 10% of all brain malignancies occur [2]. The most common tumors of this region include vestibular schwannoma, which accounts for approximately 80% of the CPA tumors [2]. Other common pathologies involved in this area include meningioma, epidermoid tumors, arachnoid cysts, lipoma, and metastases [11]. Choroid plexus papilloma (CPP) occurs very rarely in CPA and can primarily originate from choroid plexus protruded from Foramen Luschka due to direct spread of the tumor via Foramen Luschka [18,20,35].

CPA tumors may be presented by a variety of clinical manifestations due to involvement of structures in this area, such as the effect of compression on the cerebellum and cranial nerves and branches, and the effect of compression on the brain stem (including medulla and pons).

However, given the high prevalence of schwannoma in this area, the most common manifestations are vertigo, tinnitus, and hearing loss. schwannoma is commonly associated with symptoms of tumor pressure on the surrounding structures such as unilateral hearing loss, tinnitus, and vertigo [40].

Meningioma is the second common tumor of CPA with similar clinical symptoms such as hearing loss, dizziness, tinnitus, and cerebellar symptoms, but in most cases, meningioma is detected when the tumor is enlarged and compresses nerves V and X, resulting in trigeminal neuralgia, facial numbness, facial paralysis, and swallowing impairment [11,21].

Epidermoid cysts are the third most common tumors in CPA, accounting for approximately 5% of the tumors in this area [4]. Like vestibular schwannoma and meningioma, the common symptoms of these tumors are cranial nerves involvement, especially trigeminal neuralgia, hemifacial spasm, and signs of compression on the brain stem and the cerebellum [19,28].

Patients with other less common tumors, such as arachnoid cysts, have reported headache, nausea, ataxia, trigeminal neuralgia, facial numbness, hemifacial spasm, and hearing impairment [7,15].

In most cases, metastases may be confused with vestibular schwannoma; however, due to their invasive nature, the occurrence and exacerbation of symptoms of metastatic tumors in CPA are faster than other tumors [1,37].

Hydrocephalus is, by far, the most common presenting symptom for most patients with CPP [3]. The mechanism of ventricular enlargement is most commonly a combination of CSF overproduction and direct obstruction of the CSF pathways by the tumor [10]. The majority of patients present with insidious intracranial hypertension symptoms such as headaches, nausea, vomiting, and double or blurred vision [3].

The most common clinical symptoms of the patients pertained to symptoms of increased ICP, which was detected in 16 patients with hydrocephalus in radiological studies, although the symptoms of increased ICP were seen in 51 patients (Figure 1).

CSF overproduction is a rare condition and associated with two pathologies—choroid plexus hyperplasia (CPH; not a true tumor) and choroid plexus papilloma (CPP; typical or atypical)—both presenting in early childhood. Note that choroid plexus carcinoma (CPC) is not associated with CSF overproduction, probably because the cells lose their native ability to produce CSF. CPP may produce up to 5 liters of CSF per day often with a high protein level [6,13,30].

Hydrocephalus in patients with CPP can be attributed to different causes. However, unlike other areas such as ventricles in which overproduction of CSF is the major mechanism of hydrocephalus, hydrocephalus in patients with CPP in CPA is primarily obstructive due to the following reasons:

a) These tumors originate from choroid plexus and are active secretors of CSF; b) the tumor-induced pressure on the cerebellum and the fourth ventricle causes deformation of these structures and hence obstruct the CSF pathway; c) impairment in CSF absorption [13,30].

However, due to the limited CPA space and the effect of tumor compression in that area, obstruction, and deformation of the fourth ventricle seems to be the main mechanism of hydrocephalus in CPP in CPA.

Hydrocephaly in patients with CPP tumors in CPA is clinically important because, due to the mechanisms mentioned, even small size tumors can cause significant noncommunicating hydrocephalus that may even be one of the early symptoms of the illness. This is while hydrocephalus in other common tumors such as vestibular schwannoma and meningioma is often of the communicating type and occurs when

the tumor has grown to a large extent, and the other special symptoms such as hearing impairment and other symptoms associated with tumors in this area have emerged.

Particular diagnosis of a tumor in CPA is indicated by a particular combination of symptoms and imaging findings. Vestibular schwannomas, as the most frequent CPA tumor, Symptoms are often related to cranial neuropathies. Patients are more commonly present with chronic asymmetric sensorineural hearing loss confirmed by audiometry and brainstem-evoked response audiometry (>90%) and tinnitus [9]. True vertigo, unsteadiness, facial pain and numbness are unusual symptoms due to slow tumor growth [22].

As opposed to the presentation of Vestibular schwannomas, Meningiomas audiovestibular symptoms are less frequent [34]. Nevertheless, Facial pain and symptoms related to cranial nerves, which are rarely seen in VS patients, are the most common presentation of cerebellopontine meningiomas [12]. Cerebellar signs are frequently seen in these meningiomas compared with VSs, in which they are rare [16].

As mentioned before, the most common clinical symptoms of the patients with CPP in CPA pertained to symptoms of increased ICP. In addition, due to the origin of CPP tumors in CPA, i.e. choroid plexus and Foramen Luschka, even small tumors can be seen in these patients as involvement of nerves V, VII, VIII, and lower cranial nerves, alone or simultaneously, and the patients are manifested with symptoms such as hemifacial spasm, hemifacial numbness, hearing loss, and swallowing disorder [18,20,23,26].

These differences in symptoms combination probably could be helpful in differentiating CPA tumors.

# Conclusion

CPP tumors in CPA are rare but can be one of the most important differential diagnoses of adult CPA tumors, especially in cases where the patient has a combination of increased ICP symptoms along with the involvement of nerve V, hearing loss, and swallowing impairment. Frequency of symptoms is perfectly matched with the anatomical proximity to the Foramen Luschka. This variety of symptoms may not be generally observed in other common tumors of CPA, such as vestibular schwannoma or meningioma unless the tumor is very large.

# Declarations


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### Conflict of interest

The authors declare that they have no conflict of interest.

### Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the Shahid Beheshti University of Medical Sciences research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

### Informed consent

Informed consent was obtained from all individual participants included in the study.

### Authors Contribution:

Kaveh Ebrahimzadeh: Clinical management of patients, main concept of study, writing the article.

Mahmoud Omidbeigi: writing the article, review the literature, revising final edition of manuscript.

Sina Asaadi: Clinical management of patients, writing the article, review the literature, revising final edition of manuscript.

Guive Sharifi: Clinical management of patients.

Mohammad Halajnejad: Clinical management of patients, writing the article,

Mahmoud Lotfinia: Writing the article, revising final edition.

Sina Shool: Review the literature, revising final edition of article.

Omidvar Rezaei: Clinical management of patients, revising final edition of manuscript.

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# Tables

| Patient | Age/Sex | Presenting symptom | Treatment | Follow-up (Month) |
|---|---|---|---|---|
| 1 | 46/F | Hearing loss, headache, Dizziness, Decline sensation on the right side of the tongue | Complete Tumor resection | 24 |
| 2 | 32/M | Headache, Gait imbalance, Hearing loss | Complete Tumor resection | 18 |
| 3 | 42/F | Dysphagia , Gait imbalance | Complete Tumor resection | 18 |
| 4 | 41/M | Nausea, Vomiting Progressive diminution of hearing | Complete Tumor resection | 12 |
| 5 | 39/F | Headache, Dizziness, Gait disturbance | Complete Tumor resection | 18 |
| 6 | 47/F | Nausea, Vomiting, Headache, Dysphagia | Complete Tumor resection | 12 |
| 7 | 37/M | Headache, Dizziness, Tinnitus, Vertigo | Complete Tumor resection | 12 |

| | | |
|---|---|---|
| **rising** | - Hydrocephaly<br>- Increased intracranial pressure<br>- Drowsiness<br>- Progressive diminution of vision<br>- Diplopia | - Vomiting<br>- Nausea<br>- Headache<br>- Dizziness |
| **ebellar** | - Dysarthria<br>- Cerebellar sign<br>- Gait ataxia(sway) | - Vertigo<br>- Unsteady gait or unsteadiness |
| **ring sign** | - Progressive diminution of hearing<br>- Sensorineural deafness<br>- Hearing loss | - Hypoacusis<br>- Tinnitus |
| **er Motor** | - 10TH cranial nerve paresis<br>- 9TH cranial nerve paresis<br>- 6TH cranial nerve paresis<br>- Gaze palsy<br>- Optic atrophy, perception of light in both 6th nerve paresis | - Cranial nerve palsy(unknown)<br>- Dysphagia<br>- Choking<br>- Diminished gag reflex<br>- Hoarseness (laryngitis) |
| **ve 7** | - Facioplegia<br>- Hemifacial spasm<br>- Left 7th nerve paresis | - Facial weakness<br>- Loss of taste sensations on left half of tongue |
| **ve 5** | - Paroxysmal lancinating pain<br>- 5TH palsies | - Eye pain & facial pain<br>- Facial numbness |
| **hiparesis** | - Limb numbness<br>- Limb weakness | |

Table2.

Based on the previous reports and reviews, clinical symptoms of all patients were divided into seven categories.

# Figures

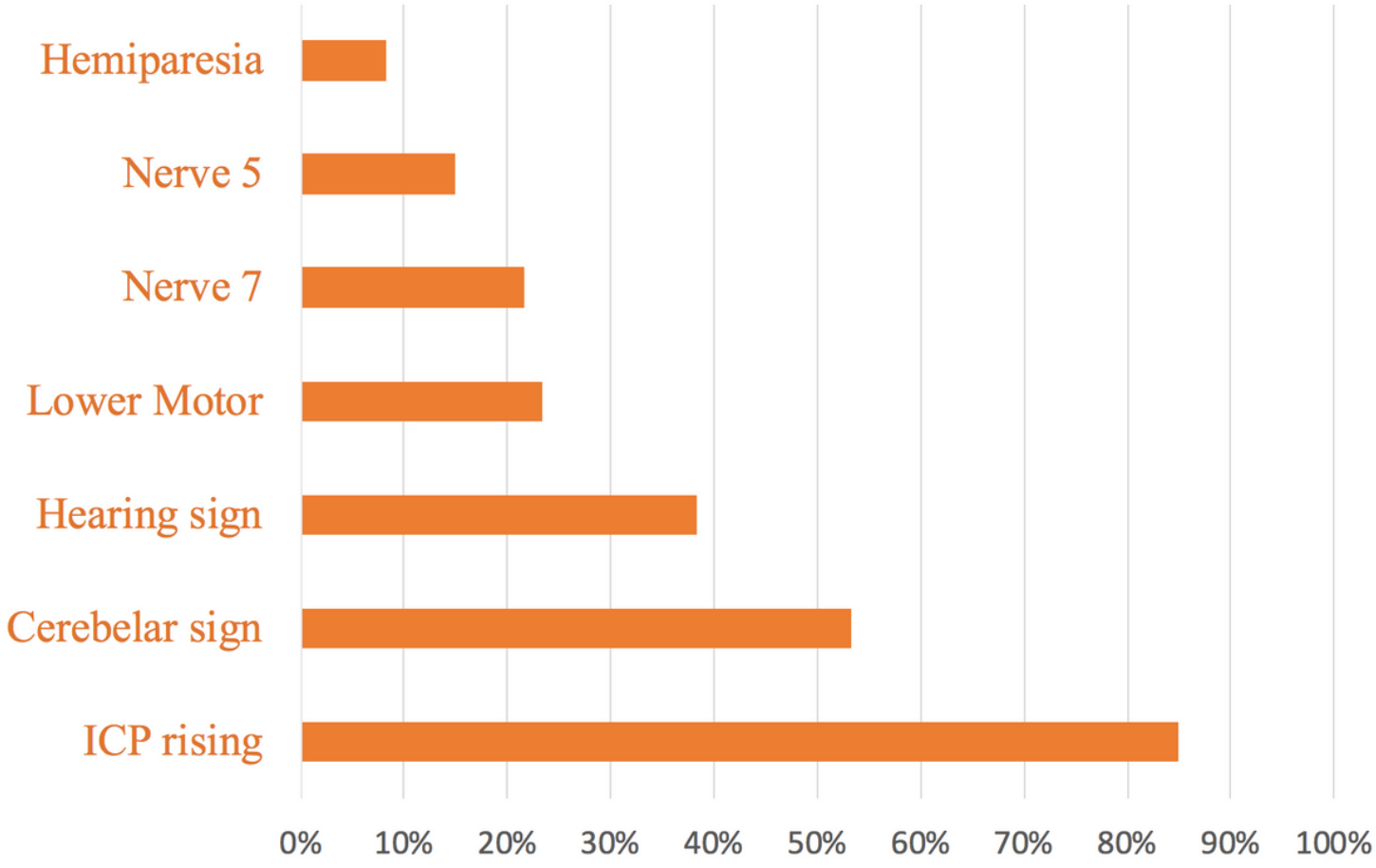
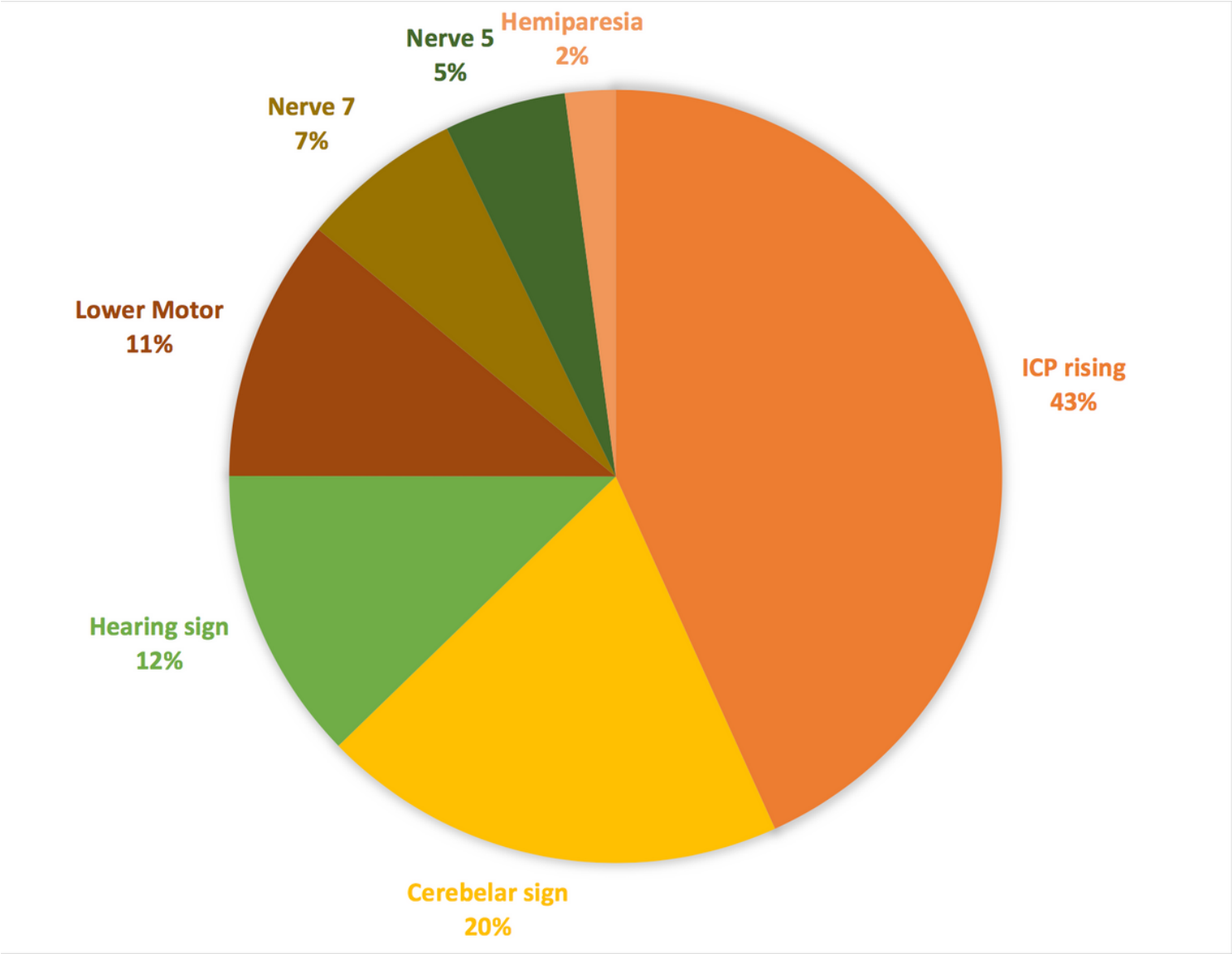



**Figure 1**

Clinical symptoms graph




## Figure 2

Clinical symptoms pie chart