Periodic abducens nerve palsy in adults caused by neurovascular compression
K A Sandvand, G Ringstad, E Kerty

ABSTRACT
Unilateral abducens nerve palsy with periodic recurrences is a well-recognised finding in children, but is rare in adults. The underlying pathophysiological mechanism is unknown. Vascular compression of the nerve is suspected but never demonstrated. We describe an adult patient with, altogether, 11 periods of unilateral right-sided abducens palsy and arterial contact at the root exit zone of the symptomatic side.

Recurred isolated lateral rectus muscle palsy is a rare, but well-recognised, finding in paediatric literature. The condition is often associated with viral illnesses or vaccination, but can occur without any known precipitating factor in previously healthy children. In some cases, structural lesions have been described, but the majority of the cases are considered to be idiopathic. Only a few cases have been reported in adults. The episodes occurred with varying time intervals, ranging from 1 week to 12 years. Most of the patients experienced painless, ipsilateral recurrences that recovered spontaneously within 6 months. No specific aetiology could be established by any of the authors.

We present a patient with recurrent right abducens palsy in whom a vascular contact to the abducens cranial nerve is documented. We believe this to be the first reported case to show, in detail, the anatomical relationship between the sixth cranial nerve and a compressing artery in recurrent, periodic abducens palsy.

CASE REPORT
In 1996, a 38-year-old man with unremarkable family history developed horizontal diplopia. Importantly, his past history showed chronic glomerulonephritis (since 1979) and well-regulated systemic hypertension.

Four months prior to his first episode of diplopia, he noticed bilateral tinnitus.

On examination, he had an esotropia of 4 degrees for near and 8 degrees for distance caused by paresis of the right lateral muscle. His visual acuity was 1.0 in both eyes, and stereo acuity tests showed suppression of the left eye. The magnitude of his abducens palsy increased gradually over a period of 2–3 weeks to a maximum of 10 degrees esotropia. This then remained stable for 6–8 weeks before improving gradually to full recovery over the following 2–3 weeks.

Seven months later, he experienced a recurrence of his right-sided abducens palsy. The clinical course was identical with the first episode and the symptoms lasted for about 3 months, again with full recovery. After that, there was a periodicity of about 7 months with only one exception; in 1999, he had undergone a kidney transplantation at the expected time of the next new episode of abducens palsy, but the next episode occurred after 21 months.

Prior to all the episodes, his tinnitus increases, he has headache and slight vertigo. During the palsy episodes, he uses glasses with 20 prisms to avoid diplopia. Between the episodes, he has no diplopia or other symptoms. The eyes are parallel with normal stereocuity; no esophoria or hypermetropia can be detected.

Recurrent neurological examinations were normal except for the periodic right abducens palsy. His episodic diplopia was present prior to and after his kidney transplantation; there is no correlation with his renal function (creatinine level) nor with his postoperative immunosuppressive medication, which are low doses of corticosteroids and cyclosporine. Under medication, his blood pressure is stable and within the normal range.

Laboratory studies showed no abnormality. Serology was negative, as were all rheumatoid factors and complement studies.

A lumbar puncture showed an opening pressure of 18 H2O cm. In addition, the cerebrospinal fluid was unremarkable. Evoked potentials VEP, SEP and AEP were all normal.

The edrophonium test was negative. He had no AchR antibodies and the single-fibre electromyography was also normal. A chest X-ray was normal.

Cerebral and orbital magnetic resonance imaging (MRI) were performed three times, once also during attack, showing small, unspecific changes of vascular origin in both hemispheres. MRI-angiography with constructive interference in steady state (CISS) sequens showed neurovascular contact between the abducens nerve and anterior inferior cerebellar artery. The diameter of the abducens nerve on the right side was smaller than the diameter of the nerve of the asymptomatic side (fig 1).

DISCUSSION
Our patient, until now, had a total of 11 episodes of right abducens palsy (fig 2).

Each episode lasted for approximately 3 months, starting slowly, reaching its maximum after 2–3 weeks. The palsy was stable about 6–7 weeks and, during this time, the patient wore eye glasses with prism with good effect. After that, he went through a recovery period of 3–4 weeks, until full normalisation of eye motility was achieved. At the start of the episodes, he had a slight headache and tinnitus. Typically, the recurrent abducens palsy is painless but, in some cases, headache was also present. The combination of headache and abducens palsy could indicate ophthalmologic migraine but, in these cases, the headache is ipsilateral to the abducens palsy, severe, and the first attack is commonly seen in

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infancy or childhood. In addition, MRI shows gadolinium enhancement of the affected nerve during attack.7 Our patient does not meet these criteria.

A few reports described isolated abducens palsy due to vascular compression but no recurrent palsy. The regularity of episodes with abducens palsy makes our case unique. The patient has had episodes approximately every 7 months, except at the time of his kidney transplantation and the interval between the two periods was almost three times the usual length.

A close relationship between the right abducens nerve and the anterior inferior cerebellar artery was detected on MRA with CISS sequence. We believe that this finding is causative in this case, similar to other syndromes involving vascular compression within the distribution of cranial nerves, such as trigeminal neuralgia and hemifacial spasm. It is generally agreed that it is only when vessels are in contact with a cranial nerve at or proximal to the root entry or exit zone that pathology results.10 The root entry zone is defined as the transition zone between the central and peripheral myelin. The central “glial” segment of a cranial nerve is covered by myelin derived from oligodendrocytes, whereas the peripheral segment of the nerve is covered by myelin derived by Schwann cells. Pathophysiologically, vascular compression syndromes are hypothesised to develop because the “junctional area between central and peripheral myelin” lacks perineurium and epineurium; therefore, it is particularly vulnerable to continued pulsatile pressure, which may result in focal demyelination and in a short-circuiting of the impulses.12 The first detailed description of ultrastructural abnormalities in the region of vascular compression was in 1994, by Hilton and colleagues.13 The authors observed focal loss of myelin and close apposition of demyelinated axons. The findings were confirmed in a subsequent electron microscope study of trigeminal specimens.14 The same authors in a later paper showed that, in the zone of demyelination, there were small numbers of thinly myelinated axons, reflecting either demyelination and remyelination or partial demyelination of the affected fibres.10 The root entry (or exit) zone is located at different distances from the brainstem for the different nerves and there is great variation in individual persons.12 The abducens nerve is a tiny structure with a diameter of 2.2 mm (range 1–2.9 mm) and an oblique course, which is the main reason for the difficulties in visualising it using conventional sequences. However, using 3D CISS, almost 100% of the abducens nerve can be identified.17

Patients suffering from trigeminal neuralgia have symptoms in periods of weeks or months, with subsequent spontaneous remission that may last for months or years. Remyelination may also be responsible for spontaneous remission of trigeminal neuralgia in some patients.15

In our case, the presence of the anterior cerebellar inferior artery compressing the abducens nerve at the root entry zone on the corresponding side might implicate neurovascular compression as the cause of the periodic palsy. Finding that the abducens nerve on the affected side was atrophic, compared with the other side, strongly suggests repeated demyelination and remyelination of this nerve.

Surgical decompression causes rapid clinical recovery both in trigeminal neuralgia and hemifacial spasm. We discussed the possibility of microvascular decompression in our patient, but the risk for complications was considered to be too high.

Competing interests: None.

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Augusta Déjerine-Klumpke (1859–1927) and her eponym

Augusta Déjerine-Klumpke was a renowned neurologist, neuroanatomist and an eminent physician of her time. She is well known for her innovative contributions to neurology and neuroanatomy. Déjerine-Klumpke was the first woman in France to accomplish entering academic life in medicine and was the first female extern and intern to practice in the hospitals of Paris. Klumpke was born in 1859 in San Francisco as the second child of John Gerard Klumpke and Dorothea Mathilda Tolle. Her mother embarked for Europe to have Augusta’s elder sister treated. During their stay in Europe, she studied in Germany and Switzerland. In 1875, her mother was informed by a fashion magazine that a woman, Madeleine Brès (1842–1921), had passed her doctorate thesis at the medical faculty in Paris. This news attracted Augusta to study medicine. However, there was no faculty of medicine in Lausanne. Therefore, the family moved to Paris since there were more opportunities for education of all of the siblings. In 1876, Augusta Klumpke started studying medicine and became a hardworking student. She studied in many laboratories and clinics. Her knowledge of foreign languages allowed her to read original papers and to consult original textbooks.

The medical society in France deemed women intellectually and physiologically incapable of making a career in medicine, but Blanche Edwards (1838–1941) and Augusta Klumpke challenged this fierce opposition. Fortunately, authorities for access for women to an externship and internship competition was given in 1882 and both women became the first female Externes des Hôpitaux de Paris. The difficulty was overcome by Paul Bert (1833–1886), a physiologist and minister of public education. The first externship years of Klumpke were spent in the service of Dr Empis. In this clinic, she diagnosed Erb’s palsy in a young patient after her chance reading of the original work of Wilhelm Erb (1840–1921).

The second and third externship years were spent in the service of Professor Edmé Félix Alfred Vulpian (1826–1887). This clinic was a general medical service, with a very large neurological consultation. During her consultations in this clinic, she made a diagnosis of a total brachial plexus radicular paralysis with oculopupillary involvement (Claude Bernard–Horner syndrome). This observation would later become known as the Klumpke type of oculopupillary involvement (Claude Bernard Horner syndrome). Pathological findings associated with the abducens nerve: surgical anatomy of its cisternal segment. Neurosurgery 1983;45:797–806.

Klumpke’s participation in her husband’s writings was important. She was a skilled illustrator and showed unrivaled dexterity when slicing microscopic preparations from the central nervous system. One of the great accomplishments of the Déjerine couple’s collaboration was two volumes of text, Anatomie des Centres Nerveux (1895, 1901).

During World War I, she made major advancements in the treatment and rehabilitation of soldiers with injuries to the nervous system, particularly spinal cord injuries. Augusta contributed to the formation of standards of assessment and treatment of these patients and also established a specialized medical and vocational rehabilitation programme.

She published numerous important papers on neurology and neuroanatomy. She also wrote a detailed autobiography written in French. In 1914, she was elected the first female president of the French Society of Neurology. Probably among her most outstanding honours, especially exceptional for a woman, was the Legion of Honour, bestowed to her first in 1915 for her scientific research and again in 1921. Klumpke will be remembered as the first female writer of a neuroanatomy textbook and for her description of the palsy resulting from injury to the inferior part of the brachial plexus. Ten years after her husband’s death, Klumpke died in 1927, at the age of 68, and was buried next to her husband in Paris at the Pére Lachaise churchyard.

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