Self-induced stretch syncope of adolescence: a video-EEG documentation

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ABSTRACT – We present the first video-EEG documentation, with ECG and EMG features, of stretch syncope of adolescence in a young, healthy 16-year-old boy. Stretch syncope of adolescence is a rarely reported, benign cause of fainting in young patients, which can be confused with epileptic seizures. In our patient, syncopes were self-induced to avoid school. Dynamic transcranial Doppler showed evidence of blood flow decrease in both posterior cerebral arteries mimicking effects of a Valsalva manoeuvre. Dynamic angiogram of the vertebral arteries was normal. Hypotheses concerning the physiopathology are discussed. *[Published with video sequences].*

Key words: syncope, stretch syncope self-induced, vertebro-basilar system, Valsalva manoeuvre, epilepsy misdiagnosis

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Loss of consciousness in syncope is caused by a transient diminution of arterial blood flow in the brain. This condition is not uncommon during adolescence and early adulthood, and is most often related to a vaso-vagal mechanism (Benditt *et al.* 2004, Wieling *et al.* 2004). Stretch syncope in adolescence (SSA) is a distinctive clinical syndrome, which is infrequent and probably misdiagnosed, and is characterized by loss of consciousness while the patient performs a stretching manoeuvre, with the neck hyperextended (Pelekanos *et al.* 1990). Its pathophysiology remains somewhat debatable (Sturzenegger *et al.* 1995). We report the first video-documented case of self-induced SSA, in which transcranial Doppler (TCD) exploration confirms a previously hypothezised physiopathology.

Case report

This previously healthy 16-year and 3-month-old boy, without any antecedents, was admitted to the adult video-EEG department of the Nice University Hospital for repeated episodes of transient loss of consciousness with falls, attributed to atonic epileptic seizures. At age 14, transient episodes of blurred vision, sometimes associated with visual flashes, occurred one to three times a month. At age 15 years and 8 months, symptomatology worsened, visual symptomatology being occasionally followed by sudden loss of consciousness and muscular tone, resulting in falls. Since then, stereotyped episodes have occurred at a frequency of two to three times per week, whereas the episodes of visual flashes occurred many

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times a day. These episodes took place predominantly during school days, but were infrequent during holidays. Clinical examination showed an athletic (180 cm tall, 82 kg) teenager with normal cardiac and neurological examination. Blood pressure was 120/80 mmHg with no orthostatic hypotension. Carotid sinus massage was negative. Baseline ECG and ECG monitoring for 24 to 48 hours, Doppler examination of extracranial carotid and vertebral arteries, and head-upright tilt-table testing were all negative. Waking, sleep, and sleep-deprived EEG were normal. Brain and cervical spine MRI showed no abnormalities of the posterior fossa or cranio-cervical junction. Because this patient's symptomatology was initially attributed to a focal occipital epilepsy, anti-epileptic treatment had been considered. Valproate, 1 250 mg a day for 4 months had no effect on the visual symptoms, or the frequency of falls. A 48-hour video-EEG was diagnostic. Twelve typical episodes were recorded, including eleven episodes of visual flashes and/or blurred vision, and two severe, atonic falls with pallor and fainting that occurred while the patient was in a standing position. All were preceded by stereotyped stretching actions combining a posterior hyperextension of the neck, with or without a lateral rotation of the head, associated with an abduction-external rotation of the shoulders. Interictal EEG and ECG were normal. During subjective visual flashes *(see video sequence 1)*, 5 to 7 seconds after stretching, the EEG showed a slowing of background rhythms that were replaced by monomorphous, middle-amplitude 4 Hz theta activity for 5 seconds. During atonic falls *(see video sequence 2)*, rapid slowing of background rhythms began 2 seconds after the end of the stretching manoeuvre and led to monomorphous high-amplitude 2.5 Hz delta waves for 5 seconds, shortly replaced by theta, then alpha activity *(figure 1)*. During stretching, the ECG showed low amplitude QRS complexes that increased after the stretch ended, suggesting the effect of a Valsalva manoeuvre.

Static and dynamic angiograms of the supra-aortic trunks revealed neither anatomical nor endovascular anomalies, apart from a spindly basilar trunk. The action of dorsal stretching carried out in a supine position then in a standing position during the second, dynamic angiogram showed no abnormalities. TCD was performed on the patient in a sitting position, using a headset fixing two transducers over the temporal areas. Two 2 MHz Doppler channels (Waki 2-Tc, Atys Medical, France) were used simultaneously. Identification of both posterior cerebral and basilar arteries was carried out according to a tech-
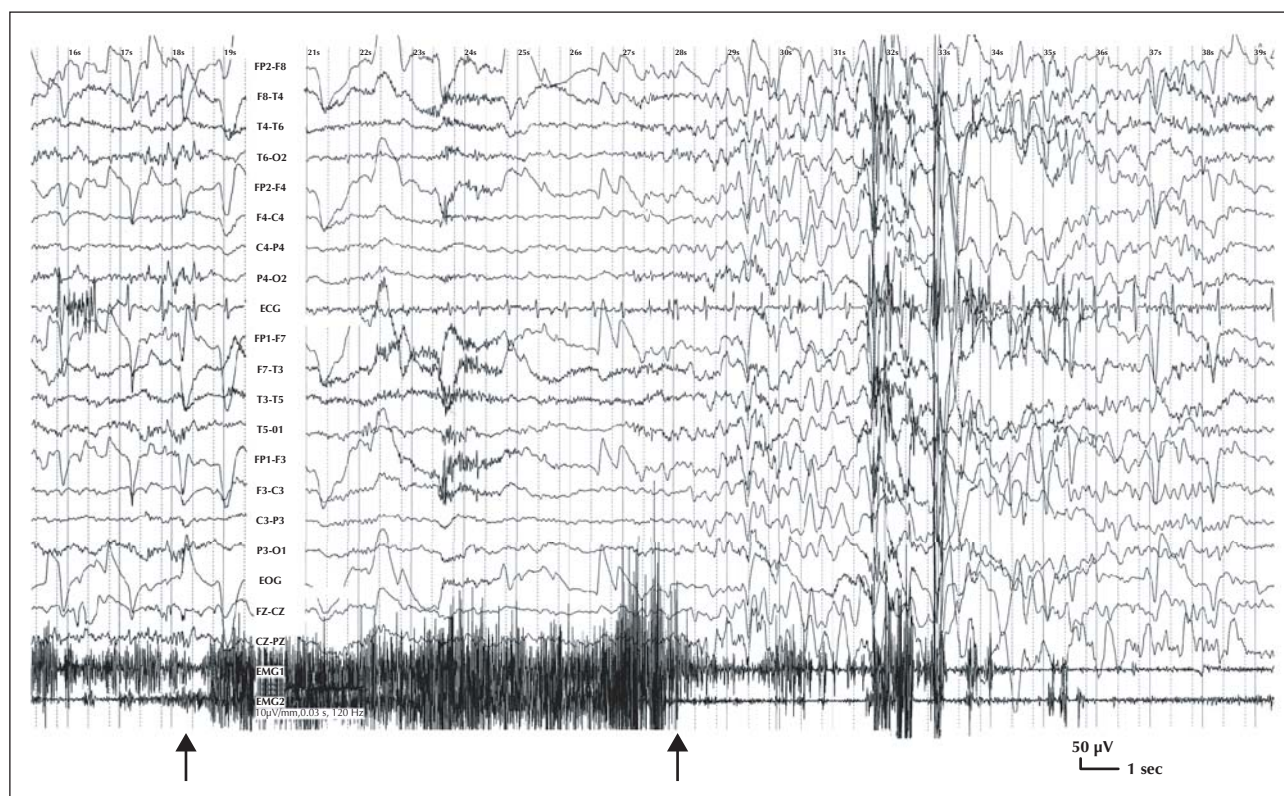

Figure 1. EEG of stretch-syncope of adolescence. The first arrow marks the beginning of the stretching manoeuvre; the second arrow marks its end. Two seconds after the end of the gross EMG artefact, a rapid slowing of background rhythms led to monomorphous high-amplitude 2.5 Hz delta waves for 5 seconds, intermixed with falling artefacts, shortly replaced by theta, and then by alpha activity. Note that during stretching, the ECG shows low amplitude for the QRS complexes that increases after the stretch ends, connected to Valsalva's manoeuvre effects.
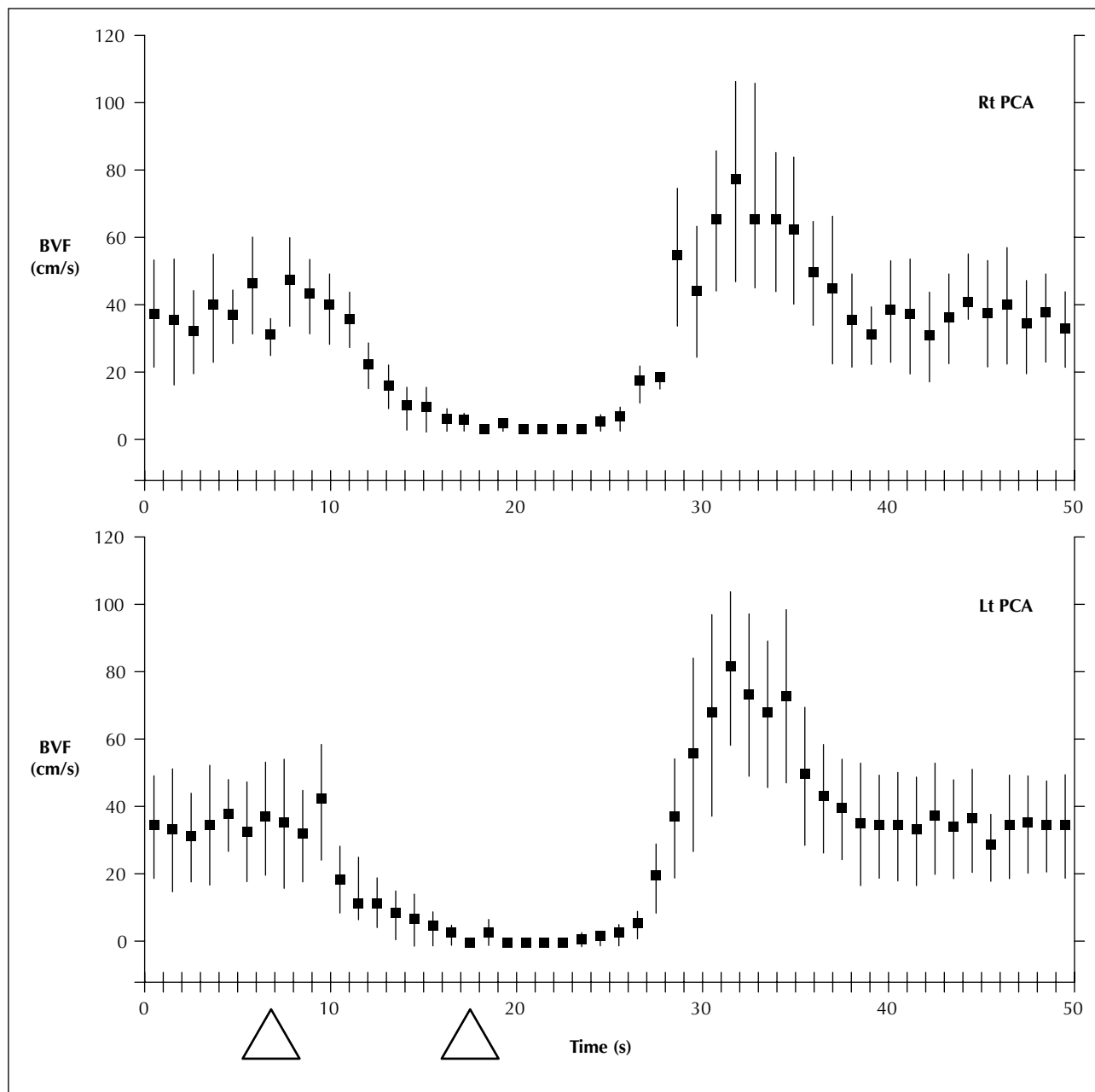
Figure 2. Time-course of blood velocities (BFV) in the right (Rt) and left (Lf) posterior cerebral arteries (PCA) when the patient was asked to perform a combination of forced hyperextension of the neck with maximum stretching, while arms were hyper-abducted. The first arrow marks the beginning of the manoeuvre; the second arrow marks the end of the inspiratory phase of the stretch manoeuvre. There is a slight increase in BFV at the onset of stretching. A fall in BFV is induced by the manoeuvre, which is then followed, 15 seconds after its termination, by an overshoot of the BFV.

nique described elsewhere (Sturzenegger *et al.* 1995). The patient was instructed to perform the following movements: 1) side rotation of the head to the right, then to the left; 2) hyperextension of the neck without stretching; 3) a combination of a forced hyperextension of the neck with maximum stretching while the arms were hyper-abducted. The procedure was repeated three times in order to con-

firm reproducibility. During a Doppler examination, blood flow remained unchanged during steps 1 and 2. With the waning of the third step, lasting almost 10 seconds, the blood flow velocities decreased significantly to below baseline in both posterior cerebral arteries *(figure 2)*, while the patient experienced his initial symptoms. When the patient returned to his initial position,

blood flow velocity returned to normal after 10-15 seconds, followed by an increase in flow to over 100% of basal value for 5 to 7 seconds that was associated with an acceleration of sinusal heart rate.

After diagnosis was confirmed, the patient was asked to explain why he did not spontaneously report that his fainting episodes were always preceded by a voluntary stretch manoeuvre. Although his answers tended to rationalize the problem, it soon became obvious that his behaviour was a way avoiding going to school. Since he stopped stretching, neither the syncope nor the visual symptoms have recurred and he continues to undertake regular sporting activities without any problem.

Discussion

This case report offers the first video-EEG description of SSA, a rarely reported, benign condition that can be mistaken for other types of syncopes or an epileptic syndrome with tonic or atonic seizures. The medical history, clinical phenomenology of the faints, and the spontaneous course of the syndrome was similar to the eight previously published cases (Pelekanos *et al.* 1990, Sturtzenegger *et al.* 1995), except that our patient consciously induced the triggering manoeuvre to avoid going to school. Syncopal attacks, including those compulsively induced by the Valsalva manoeuvre, may also induce genuine epileptic sei-

zures (Aicardi *et al.* 1988, Stephenson *et al.* 2004, Horrocks *et al.* 2005), adding to the diagnostic difficulties. However, although one of the adolescents with stretch syncope in the study by Pelekanos *et al.* (1990) was reported to have had an anoxic seizure, epileptic seizures have not been described following stretch syncope. Since our patient was told not to stretch in his usual manner, he has remained free from any kind of neurological disorder.

SSA is a distinctive clinical syndrome, which involves complete loss of consciousness, whilst in a standing or sitting position after stretching while the neck is hyperextended (Pelekanos *et al.* 1990). This rarely reported type of syncope occurs predominantly in boys. This is contrary to vaso-vagal syncopes, which are more frequent in girls (Benditt *et al.* 2004, Wieling *et al.* 2004). Study of the cardiovascular responses to stretching indicates that a Valsalva manoeuvre cannot be the only mechanism operating in these patients (Pelekanos *et al.* 1990). In all published cases, static angiogram of the vertebro-basilary system, showed no lesion. In a single case report (Sturzenegger *et al.* 1995), dynamic angiogram showed some evidence of a hemodynamic problem, with delayed filling of a vertebral artery trunk during stretching, whereas TCD monitoring of both posterior cerebral arteries in two patients showed a significant decrease in blood flow velocities when the neck was hyperextended. Similar findings were documented in our patient, supporting the hy-
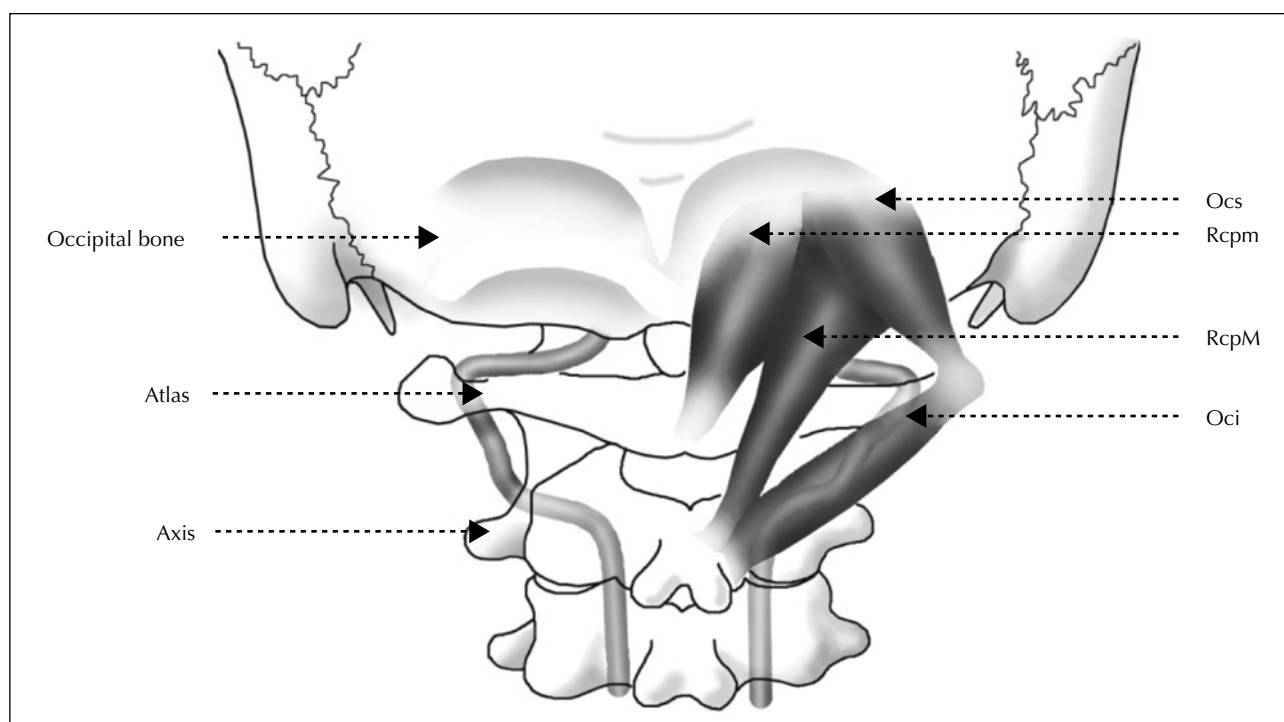

Figure 3. Anatomical relationship between the vertebral artery and the deep muscular plane of the neck.
Rcpm: muscle rectus capitis posterior minor; Ocs: muscle obliquus capitis superior; RcpM: muscle rectus capitis posterior major; Oci: muscle obliquus capitis inferior.

pothesis of a transitory, functional brainstem ischemia at the vertebral artery level. Mechanical compression of the extracranial atlas loop segment of the vertebral arteries during neck rotation may also be investigated by TCD in elderly patients with evidence of unexplained, positional, vertebrobasilar-distribution symptoms (Sakaguchi *et al.* 2003).

However, because of normality of the patient's dynamic angiogram and based on the TCD findings, the mechanical effect of the stretching manoeuvre may also have induced the same effects on the arterial blood pressure as the Valsalva manoeuvre. Unfortunately, we cannot provide continuous recording of our patient's systemic blood pressure during the stretch manoeuvre. However, the blood flow deceleration observed during the dynamic TCD in the vertebral artery could not be due only to Valsalva's manoeuvre effect. Considering the anatomical variations of this area (Rocha *et al.* 2007), we hypothesised that a peculiar anatomical background may exist *(figure 3)*. Hyperextension of the neck may allow the deep muscular plane of the neck consisting of the *rectus capitis posterior minor* and the *rectus capitis posterior major* muscles to be in contact with the horizontal portion of the vertebral artery (Rutten *et al.* 1995). This portion of the artery may be trapped between these muscular masses and the atlanto-occipital posterior membrane. Abduction of the upper arm

may induce contraction of the superficial muscular plane made up of the *trapezoid* muscle, which compresses the underlying muscle plane, and as a result enhances the flux reduction of vertebral arteries *(figure 3)*. Because of the rarity of diagnosed SSA, conducting anatomical studies to demonstrate the impact of this potential anatomical predisposition will be difficult. In conclusion, both the performance of a Valsalva-like manoeuvre and compression of the vertebral arteries during the stretching manoeuvre appear to be involved in SSA, but the relative roles of these two factors remains to be elucidated. Other factors such as genetic, neurally-mediated syncope may also be involved. □

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Legends for video sequences

Sequence 1: video-EEG during subjective visual symptoms induced by the characteristic stretching manoeuvre. Five to 7 seconds after stretching, the EEG shows a slowing of background rhythms, which are then replaced by monomorphous, middle-amplitude 4 Hz theta activity for 5 seconds, while the patient is slightly obtunded for 2 seconds.

Sequence 2: video-EEG of a full syncopal episode shortly after the stretching manoeuvre was performed in a standing position. During the fainting episode with atonic fall, rapid slowing of background rhythms begins 2 seconds after the end of the stretching manoeuvre, and is followed by monomorphous high-amplitude 2.5 Hz delta waves for 5 seconds, shortly replaced by theta, then by alpha activity. A second sequence in slow motion follows the first one, and shows upward, eyeball deviation immediately preceding the atonia.