

SPONTANEOUS ATLAS DISLOCATION¹

By

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As far as is known, spontaneous dislocation of the atlas without demonstrable skeletal destruction was described for the first time by *Bell*, 1830. Since then fully a hundred cases have been published, often listed under the heading of acquired torticollis.

Cases appearing in the literature were collocated in 1932 and 1934 by *Watson-Jones* who added 6 cases of his own, in 1949 by *Sullivan*, who added 4 cases, and 1953 by *Boever & Hennebert* with the addition of 6 cases. A number of authors like *Grisel*, *Burgeois*, *Berkheiser & Seidler*, *Gillette*, *Fitzwilliams*, *Grogono* etc. have published two or more of their own cases.

According to several articles varying forms of dislocation may occur, but only the most common form will be discussed here; this manifests itself through a unilateral forward anterior luxation in one of the intervertebral joints between the atlas and the epistropheus. The other forms of dislocation are probably only of theoretical interest.

Nearly all case histories reveal acute or subchronic inflammatory processes within the upper region of the throat or the region at the base of the skull, preceding the dislocation by a period varying from a day up to several months.

The clinical picture of a spontaneous atlas dislocation is simple in the uncomplicated cases. A patient appears with torticollis to a more or less marked degree. The deformity does not differ from that customary in congenital torticollis. The contracture or fibrosis typical of congenital torticollis in the sternocleidomastoid muscle is lacking, however, although a contracture may sometimes be demonstrated in the neck musculature. If the dislocation of the atlas is extreme, a bulge will be found in the retropharynx, owing to the projecting atlas arch. An olithesy depression can be demonstrated in such cases in the neck hollow. In lesser dislocations the torticollis position is not so pron-

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unuced and is certainly often overlooked. What is obvious however is the manner in which a patient observes his surroundings, by avoiding turning his head in the one direction, depending on which side is dislocated (Fig. 2 b). The face can be turned away from but not towards the dislocated side (Figs. 2 a-c).

In the differential diagnosis, apart from congenital torticollis, various forms of muscle contractures, skeletal affections and anomalies may call for consideration.

The final diagnosis is made by means of the roentgen examination. On the frontal picture the intervertebral joint fissure of the dislocated side is revealed as having disappeared owing to overlapping, which arises on dislocation (Fig. 4 b). On the lateral picture an increased distance is present between the anterior atlas arch and dens epistrophei (Figs. 1-4). This increased distance between the anterior atlas arch and dens epistrophei may also be clearly observed in axial pictures of the cranium.

The pathophysiology of the dislocation has been discussed by *Watson-Jones*, amongst others, who considers that the dislocation is due to a hyperaemic decalcification of the attachment of the transverse atlantal ligament, brought about by adjacent inflammation, and followed by a total or partial rupture. The extent and consequences of the dislocation should be in direct proportion to the extent of the rupture. Another opinion is represented by *Grisel*, who considers that the primary factor in the pathogenesis is a muscle contracture, determined by an inflammation of the attachments of the deeper neck muscles. As a result of this contracture the atlas would be pressed forward, "enucleated", to its dislocated position.

Isolated cases with lethal results have been published (*Bell, Watson-Jones, Ramond, Apert*). In these cases death was caused by a compression of the medulla oblongata. Serious complications in the form of reflex disturbances, sensibility disturbances, pareses and paraplegias occur relatively often amongst the published cases.

A very good prognosis is given for the uncomplicated cases, as a rule with recovery. Even cases with neurological symptoms are considered to have a good prognosis. *Berkheiser & Seidler* amongst others recorded a case with quadriplegia which gained full recovery. On the other hand *Ramond* gave an account of a case with quadriplegia which led to death after a day.

The treatment consists in reduction by means of permanent traction with Crutchfield tongs, Glisson sling or a similar arrangement or by reduction under narcosis followed by plaster bandage. Fixation for about three weeks is usually considered to be satisfactory. In some



Fig. 1 a.

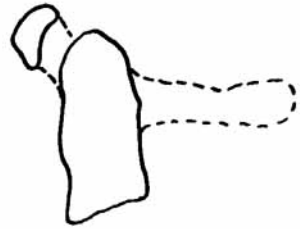
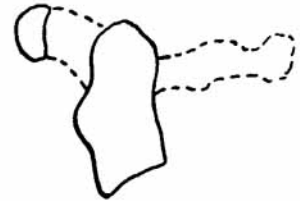


Fig. 1 b.

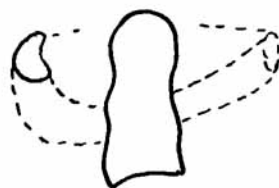


cases, however, the displacement is liable to recur, and permanent traction is recommended in such cases for a fairly long period with the head kept in a dorsally-flexed position, followed by a plaster neck jacket for a period of three monthes. For such cases *Grogono* suggests operative fixation of the posterior atlas arch to the spinous process of the epistropheus.

CASE REPORTS

A case with a one year history of acquired torticollis was referred to the orthopedical out-patient department in Lund for diagnostic clarification. This occasioned a study in the literature bearing in the first place upon treatment and prognosis since the case seemed to be on the point of developing serious complications.

It was obvious that the number of published cases was relatively small but that authors often published several personal cases. This

*Fig. 1 c.**Fig. 1 d.*

brought on the reflexion that such atlas dislocations were in all probability not so rare, but that they were overlooked for one reason or another. A deliberate search for such cases produced a number with acquired torticollis and amongst these the following four undoubtedly belong to the above type.

Case 1. A girl aged 4. Fig. 1 (a-g).

Earlier always healthy. No known trauma. She went for a boat trip with her parents one Sunday in May, 1952. On the following day there was malposition of the head and she felt pains in her neck on incautious movements. Gradually a rheumatological polyarthrititis picture developed and the patient was admitted to the rheumatological clinic in Lund, 1953, from which she was sent in March, 1953, to the orthopedic out-patient department because of the painful malposition of the head. She then used both hands to support the head when changing

*Fig. 1 e.**Fig. 1 f.**Fig. 1 g.*



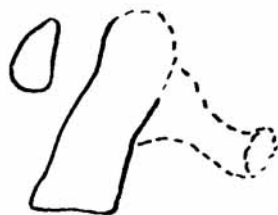
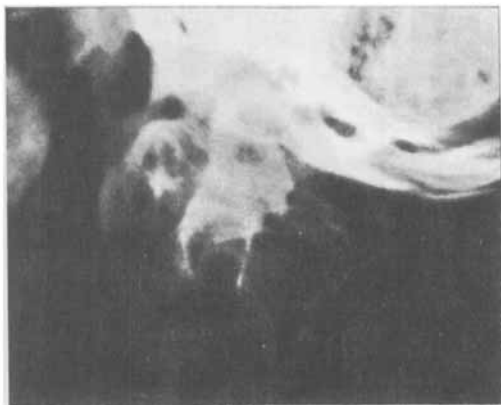
Fig. 2 a.



Fig. 2 b.



Fig. 2 c.

*Fig. 2 d.*

position and manifested a torticollis deformity. There was pronounced muscle contracture and tenderness in the neck region. The roentgen examination revealed a considerable dislocation of the atlas. (Figs. 1 c, e and f).

She was treated with extension and fixation until February, 1954. Afterwards she had no subjective trouble. There was some residual torticollis deformity, considerable limitation of movement in the spinal column at the neck and partial residual dislocation of the atlas (Figs. 1 d and g).

Case 2. A girl aged 8. (Figs. 2 a-d).

Had polyarthrititis since December, 1950. She was tended at various hospitals after that time. She declares that the neck deformity arose some time during 1952.

The case was discovered when the patient was admitted to the orthopedical department in Lund owing to flexion contractures in the knee joints, March 1954. There were no residual subjective troubles apart from limitation of movements.

She was given extension treatment for two months, employing chin halter, without demonstrable effect.

Case 3. A girl aged 13. (Figs. 3 a-b).

Polyarthrititis since the age of 1 year. In 1949 or 1950 a torticollis arose.

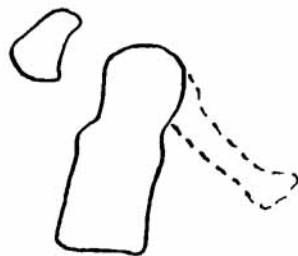
There were no residual subjective troubles apart from the malposition of the head.



Fig. 3 a.



Fig. 3 b.



Case 4. A girl aged 11. (Figs. 4 a-b).

Had polyarthrititis since 1947. She had noticed since 1953 that she found it difficult to move her head. Otherwise there were no subjective troubles.

DISCUSSION

The hyperaemic decalcification of the attachments of the transverse atlantal ligament, presupposed by *Watson-Jones*, probably requires a certain time for its accomplishment, one or several weeks, while several of the published cases have an anamnesis of only one day. Moreover the degree of dislocation could not be reasonably explained with the



Fig. 4 a.

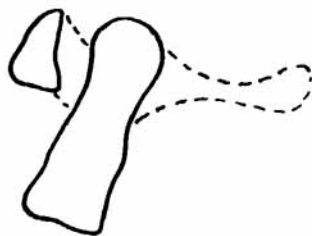


Fig. 4 b.



theory advanced by *Watson-Jones*. Partial ruptures of the transverse ligament would probably not lead to dislocation at all. Total rupture or weakening of the ligament throughout the whole of its transverse section must be presumed before a dislocation can arise. The extent of the dislocation therefore could not be assumed to depend upon partial ruptures either. The explanation given by *Grisel* does not in the first place cover the reason for the insufficiency of the transverse atlantal ligament. It is not a question either of any "enucleation" of the atlas since the relationship with the base of the skull remains unchanged. It does not seem very likely that the dislocation is determined by muscle contracture since the only muscle which could have a forward-dislocating effect on the cranium and thus the atlas is the sternocleidomastoid muscle, which however is not affected by the contracture.

These and similar problems related to the treatment of the first case brought about an investigation to ascertain the functioning of the ligamental apparatus of the neck joint, an account of which will be given later.

So much should however already be clear now that in these dislocations "life insurance" is maintained by the odontoid ligaments. The degree of dislocation probably depends on individual variations in the course and length of these ligaments.

Finally, as far as the length of the fixation treatment is concerned, the time stated—three weeks—is probably enough in insufficiencies of the transverse ligament arising in connection with some acute affection. When a complication occurs in a disease with a connective tissue affection of the chronic polyarthritis type, a longer period of fixation, possibly a longer period of bed rest with dorsally flexed head, must be considered as indicated.

As will appear from the cases described here the prognosis of inveterate cases is by no means satisfactory. It is important to stress therefore that these cases should be diagnosed at an early stage, i.e., while reduction can be achieved and healing of the transverse ligament can be expected.

Operative measures ought to be reserved for cases of reduction, which recur after the cessation of the fixation treatment.

S U M M A R Y

In March, 1953, the author received a case of spontaneous atlas dislocation for diagnostic clarification. By studying the literature it was seen that a number of cases had been published previously. The prevalent view of the dislocation's mechanism, however, could not be accepted, and this induced the author to begin an investigation, the result of which will be published later.

An assumption that such dislocations are more common than was previously supposed was verified by investigating available child patients whereupon a further three cases were revealed.

The acute consequences of an atlas dislocation owing to insufficiency of the transverse atlantal ligament is, in the author's opinion, dependent on individual variations in the length of the odontoid ligaments.

The treatment and prognosis are discussed and it is pointed out that the prognosis of inveterate cases is by no means satisfactory and therefore the importance of early diagnosis and treatment is emphasised

RESUME

Un cas de dislocation spontanée de l'atlas a été soumis à l'auteur en mars 1953 pour en établir le diagnostic.

En revoyant la littérature, il a constaté qu'un certain nombre de cas avaient été publiés antérieurement. Il ne put cependant accepter la conception habituelle du mécanisme de la dislocation, ce qui a amené l'auteur à entreprendre un travail qui sera publié ailleurs.

La supposition que ce genre de dislocation est plus courant qu'on ne l'avait présumé antérieurement s'est trouvée vérifiée par l'examen d'une clientèle d'enfants parmi lesquels il a pu constater encore trois cas.

Les suites aiguës de la dislocation de l'atlas due à l'insuffisance du ligament transversal sont causées, de l'avis de l'auteur, par des variations individuelles dans la longueur des lig. alaria.

Le traitement et le pronostic sont discutés et, étant donné que le pronostic des cas invétérés n'est absolument pas satisfaisant, il est relevé combien il est important d'établir précocement le diagnostic et le traitement.

ZUSAMMENFASSUNG

Der Verfasser hat im März 1953 einen Fall von Spontanverrenkung des Atlas zur diagnostischen Aufklärung erhalten. Bei Durchsicht der Litteratur erwies es sich, dass eine Anzahl von Fällen früher veröffentlicht worden waren. Die gewöhnliche Auffassung des Mechanismus der Verrenkung konnte er jedoch nicht anerkennen, ein Umstand, der ihn veranlasste, mit einer Arbeit zu beginnen, die später veröffentlicht werden wird.

Die Annahme, dass derartige Verrenkungen häufiger auftreten als man ehemals vermutete, bewahrheitete sich, indem beim Durchuntersuchen des zugänglichen Kindermateriales drei zusätzliche Fälle nachgewiesen werden konnten. Die akuten Folgen einer Atlasverrenkung auf der Basis einer Insuffizienz im ligamentum transversum atlantis beruhen nach der Meinung des Verfassers auf individueller Verschiedenheit in der Länge der Ligamenta alaria. Die Behandlung und Prognose wird besprochen, wobei hervorgehoben wird, dass die Prognose in veralteten Fällen keineswegs zufriedenstellend ist. Die Wichtigkeit frühzeitiger Diagnose und Behandlung wird deshalb unterstrichen.

REFERENCES

- Boever, F. & Hennebert, P.*: Des dislocations non traumatiques de la colonne cervicale. *Rev. de Chir. Orth.* 39: 24-69, 1953.
- Berkheiser, E. J. & Seidler, F.*: Nontraumatic Dislocations of the Atlanto-Axial Joint. *J.A.M.A.* 96: 517-523, 1931.
- Grisel, P.*: Énucléation de l'atlas et torticollis naso pharyngien. *La Presse Médicale* 1930: 50-53.
- Grogono, B. J. S.*: Injuries of the Atlas and Axis. *J. Bone & Joint Surg.* 36 B: 397-410.
- Sullivan, A. W.*: Subluxation of the Atlanto-Axial Joint: Sequel to Inflammatory Processes of the Neck. *J. Pediat.* 35: 451-464, 1949.
- Watson-Jones, R.*: Fractures and Joint Injuries. Edinburgh 1943.
- Spontaneous Hyperaemic Dislocation of the Atlas. *Proceedings of the Royal Soc. of Med.* 25: 586-590, 1931-1932.
 - & *Roberts, R. E.*: Calcification, Decalcification and Ossification. *Brit. J. Surg.* 21: 461-499, 1934.