

Ment. Dis., 1898, xxv, 81; *Univ. Penn. Med. Bull.*, January, 1905. [4] BINWANGER, *Virchow's Arch. f. Path. Anat.*, 1882, 427. [5] GERLICH, *Arch. f. Psych.*, 1891-2, xxiii, 203. [6] OTTO, *Arch. f. Psych.*, 1891-2, xxiii, 153. [7] HAUSHALTER and COLLINS, *Compt. rend. de la Soc. de Biol.*, 1905, lix, 223. [8] RHEIN, J. H., *Journ. Nerv. and Ment. Dis.*, 1913, xl, 639. [9] VOGT, C., *Jahrb. f. Psych. und Neur.*, xviii, 293, 478, 489, 1911. [10] ANTON, *Jahrb. f. Psych.*, 1895, xiv, 141. [11] BATTEN, *Brit. Journ. Child. Dis.*, 1915, xii, 65. [12] SPENCER, H. R., *Trans. Obst. Soc.*, 1891, xxiii, 270. [13] CROTHERS, B., "Birth Injuries of the Central Nervous System," Pt. II, Baltimore, 1927. (Full bibliography). [14] GREENFIELD and HOLMES, *Brain*, xlviii, Pt. II, 183. (Bibliography). [15] CROTHERS, B., VOGT and ELEY, *Amer. Journ. Child. Dis.*, xl, 227-246. [16] GUTTMAN, *Fortschr. a. d. Geb. d. Röntgenstrahlen.*, 1929, xl, 965-978. [17] STUTSMAN, RACHEL, "Mental Measurement of Pre-School Children," The World Book Company, G. G. Harrap and Co. [18] "Psychological Tests of Educable Capacity," H.M. Stationery Office.

Spontaneous Hyperæmic Dislocation of the Atlas.

By R. WATSON JONES, F.R.C.S., M.Ch.(Orth.).

ONE day in the course of her round, a nurse was dressing the sinuses in the neck of a girl aged 16 suffering from tuberculosis of the cervical glands with secondary pyogenic infection. In order to gain better access, the nurse pushed the patient's cheek with the back of her hand as she asked her to turn her head—the girl choked, became cyanosed, and was dead. Post-mortem examination revealed a forward dislocation of the atlas with compression of the cord by the odontoid process of the axis. This case, perhaps the most dramatic of its type, has been reported by Reid [1], and during the last few years, 12 other examples of spontaneous dislocation or subluxation of the atlas have been recorded by Swanberg [2], Berkheiser [3], Seidler, Grisel [4], Desfosses [5], and Grieg [6]. Although the condition is apparently little known and seldom recognized, it is just 101 years since Sir Charles Bell [7] described the case of a man with a deep ulcer on the back of his throat who sustained a dislocation of the atlas; in the words of Bell, "the ulcer had destroyed the transverse ligament of the atlas."

The condition is apparently rare, and this paper has been based on the fourteen previously recorded cases, as well as on the two cases which have occurred in my own practice and have been personally observed.

Ætiological factors.—It is of importance to note that the condition is almost confined to the first decade of life. In only three of the sixteen cases have the patients been older than 11, and the common ages are 8, 9 and 10.

AGE INCIDENCE. (ANALYSIS OF RECORDED AND PERSONAL CASES.)

Age	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22
Cases	...	1	1	2	4	2	1	2	1

SEX INCIDENCE.

Females 8 Males 6 Unknown 2

On no occasion has there been any real traumatic factor. As a rule the onset has been absolutely spontaneous, and the dislocation has been produced by a normal movement of the head. In Swanberg's case the dislocation had first appeared some hours after the operation of tonsillectomy, but in one of my cases it appeared four days before operation for acute mastoid disease.

The one predisposing factor which has been constant is the existence of an inflammatory focus in the upper neck. Grisel and Desfosses thought that the infection was necessarily nasopharyngeal in origin, and have therefore coined the term, "nasopharyngeal torticollis." More detailed study shows, however, that any infection which would cause hyperæmia in the region of the base of the skull may give rise to the condition—tonsillitis, pharyngitis, middle-ear disease, mastoid disease, inflamed cervical glands and so on. The type of organism responsible is of no significance—the infection may be pyogenic, tuberculous or syphilitic. Moreover, there need not be any direct association between the focus of infection and the atlas.

RECORDED CASES OF SPONTANEOUS DISLOCATION OF THE ATLAS.
CLASSIFICATION OF PREDISPOSING CAUSES.

Acute tonsillitis	4	Acute suppurative mastoiditis ...	1
Influenzal cold (nasopharyngitis) ...	3	Tuberculous cervical adenitis ...	1
Sore throat (pharyngitis)	2	Pyogenic cervical adenitis	1
Scarlet fever with pharyngitis	1	Cervical abscess (? retropharyngeal) ...	1
Syphilitic ulceration of pharynx	1	None known	1

Clinical features.—The history usually given is that the child was not well and had a cold or a sore throat. A week or ten days later for no apparent reason he suddenly complained of a “crick in the neck” and the mother noticed that his head was twisted and that movements were avoided. The head is held rigidly, slightly in front of its normal plane, tilted towards one shoulder, with the face rotated towards the opposite shoulder. It is, in fact, the ordinary position of any torticollis, but there is no contracture or spasm of the sterno-mastoid; the rigidity is apparently due to spasm of the deep cervical muscles. Any attempt to move the head is strongly resisted, and very much resented by the child, who moves cautiously, and will support his chin on his hands when rising from the recumbent position, or performing any movement which demands flexion of the neck. There is no tenderness over the cervical vertebræ, but percussion gives rise to pain. Skiagrams show a forward displacement of the skull and atlas on the axis. The odontoid process may lie as much as half an inch behind the anterior arch of the atlas and appears to be situated in the middle of the foramen magnum. In none of the cases which survived was there any evidence of an irritative lesion of the cord, although radiological examination makes it clear that the cord is lying in a very restricted space, between the odontoid and the back of the atlas and foramen magnum. It is clearly only a question of degree, the matter of a mere millimetre, which is separating life from death.

One feature of the greatest significance is that the anterior arch of the atlas may show marked decalcification. In the first of the author's cases, the pre-operation skiagram (fig. 1a) shows that about half of the anterior arch has completely disappeared, and if it were not for the subsequent skiagram one would suspect either a congenital anomaly, or actual bony destruction by tuberculous or pyogenic disease. The second skiagram, however, taken only three months later (fig. 1b) shows that the arch is once more intact, and its apparent disappearance has been due simply to decalcification.

Pathology.—Four important facts stand out clearly from the ætiological and clinical features.

(1) The condition is peculiar to children, at an age when the bones are imperfectly developed and not yet well calcified. (2) It is necessarily associated with an inflammatory lesion which may be situated anywhere in the upper cervical area. (3) There is a latent period of about from seven to ten days between the onset of infection and the dislocation. (4) The anterior arch of the atlas shows decalcification but not destruction.

The relationship of calcification of bone to its blood supply was not clearly understood until Grieg [6] of Edinburgh published his investigations. To him we owe the proof that hyperæmia causes decalcification of a bone, and reduction of the blood supply causes sclerosis. With this fact established, it is easy to appreciate the sequence of events in spontaneous dislocation of the atlas. The focus of infection is responsible for an ever-widening area of hyperæmia. If the focus is anywhere in the upper cervical area the field of hyperæmia will eventually include the upper cervical vertebræ and particularly that part of the atlas which is nearest to the infection. In pharyngeal infections this is the central part of the anterior arch of the atlas, and in mastoid infections the lateral part of the arch of the atlas on the same side as the mastoid (fig. 1). Decalcification is now inevitable. After a latent period of from seven to ten days in an average case of pharyngeal or tonsillar infection,

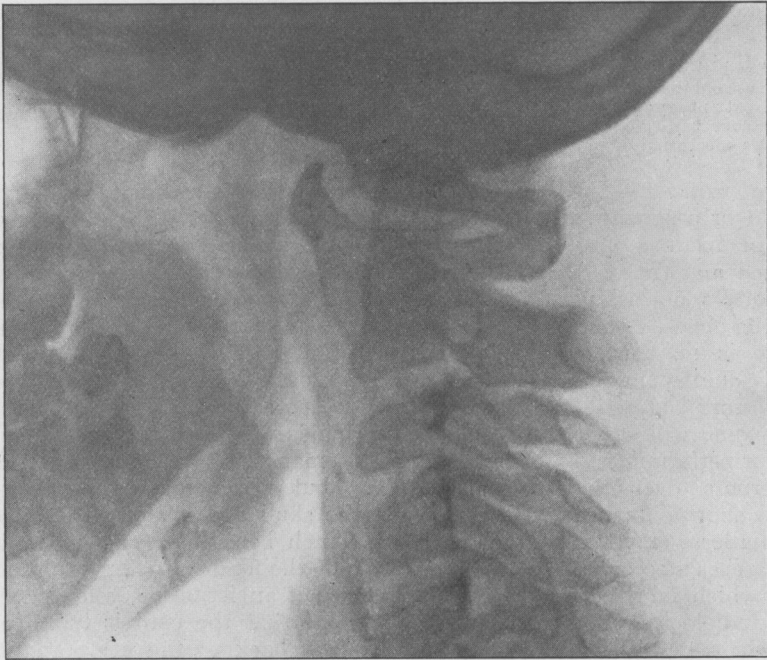


FIG. 1A.—Spontaneous dislocation of the atlas in a boy aged 9, five days after onset of symptoms of acute mastoid disease. Note that half of the anterior arch of the atlas has "disappeared" owing to hyperaemic decalcification.

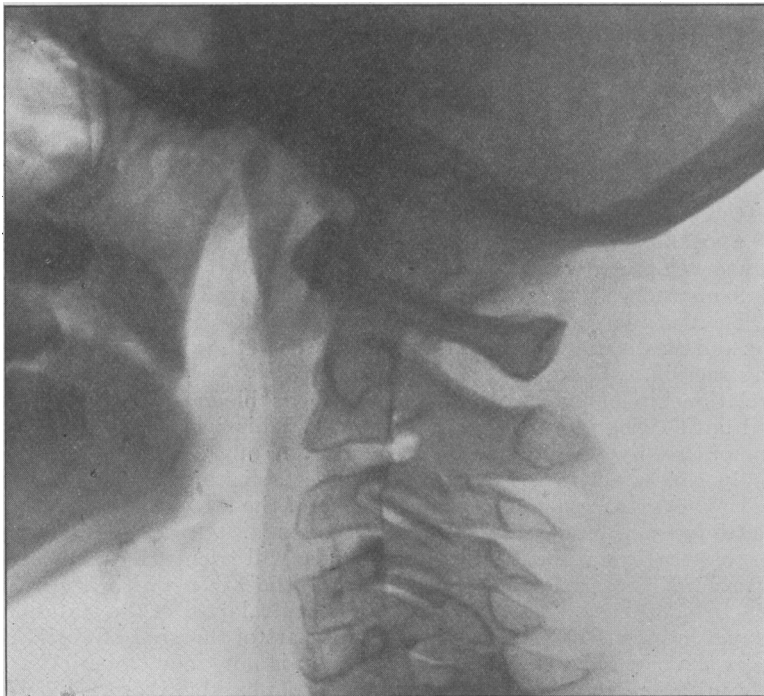


FIG. 1B.—Same case as fig. 1A, four months later. Dislocation is now fully reduced and the atlas is completely re-calcified. There was perfect recovery with a normal range of movement of all joints.

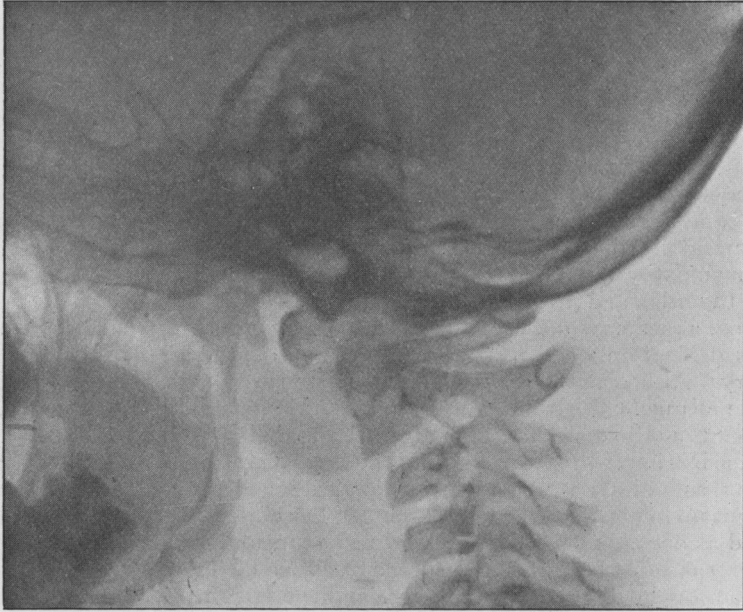


FIG. 2A.—Spontaneous dislocation of atlas in a child aged 2. In this case the hyperæmia was secondary to a simple "cold in the head" with pharyngitis.

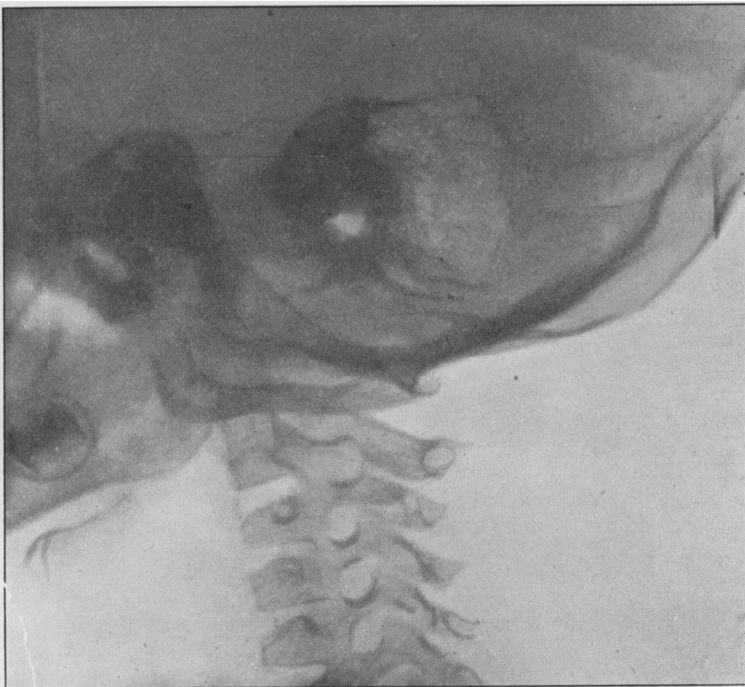


FIG. 2B.—Same case as Fig. 2A. Complete recovery in 4 months.

the degree of decalcification is such that the arch of the atlas no longer affords a secure attachment for the transverse ligament, which is mainly responsible for maintaining the odontoid process in close relationship with the atlas. A normal movement of the head may avulse the ligament and allow forward displacement of the atlas. According to the degree of avulsion, the dislocation is complete and the patient dies suddenly, or is incomplete and the syndrome we have described is established (figs. 1A, 2A).

Treatment.—Recorded cases have been treated, as a rule, by recumbency and head extension, and in many cases prolonged fixation has been deemed necessary. Berkheiser and Seidler maintained traction and then fixation for as long as twelve months, presumably because they considered the condition to be due to an infective process in the atlas and atlo-axoid joints.

Knowing, as we now do, that the dislocation is purely mechanical in origin and due to decalcification of the atlas, it is clear that prolonged fixation is not only unnecessary but not advisable, since it will give rise to still further decalcification by atrophic change.

Provided that the dislocation is not of long standing, it may be reduced with the greatest ease by simple hyperextension of the head (figs. 1B, 2B). Under an anæsthetic the muscle spasm disappears, the neck is fully extended and the head, neck, and upper trunk are put up in plaster. It is found that all discomfort, pain and fear of movement are relieved at once, and the patient need not be recumbent for more than 24 hours. Energetic steps must be taken to secure quiescence of the infection, and a sufficient time allowed for the hyperæmia to subside and the atlas to recalcify. In both of my cases the plaster was retained for 10 weeks. After re-educative exercises for two months a full recovery was made—there was a normal range of movement of all joints, no deformity, no pain and the X-rays showed complete recalcification.

REFERENCES.

- [1] REID (quoted by Grieg, 6). [2] SWANBERG, *Journ. Amer. Med. Assoc.*, 1919, lxxii, 107-8. [3] BERKHEISER and SEIDLER, *Journ. Amer. Med. Assoc.*, 1931, xcvi, No. 7, 517. [4] GRISEL, *Presse Méd.*, 1930, iv, 50. [5] DESFOSSES, *Presse Méd.*, 1930, No. 70, 1179. [6] GRIEГ, "Clinical Observations on the Surgical Pathology of Bone," 1931. [7] BELL, "The Nervous System of the Human Body," 1880, app., cxxvii, No. lxiv, 64.