Paraplegia (1974), 11, 263-267

CONGENITAL ABSENCE OF THE DENS OF AXIS

A Case Report with Tetraplegia

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EMBRYOLOGICAL AETIOLOGY

Ossification of cartilaginous of the dens of axis begins at the 5th month of foetal life (Freiberger *et al.*, 1965). Generally it develops from three ossification centres, two lateral and one apical. During the 3rd and 4th year the dens unite with the body of the axis (Rowland *et al.*, 1958). Congenital anomalies of dens are: separation of the apical or ossicle, partial and complete absence of dens (Gillman, 1950; Sherk, 1967).

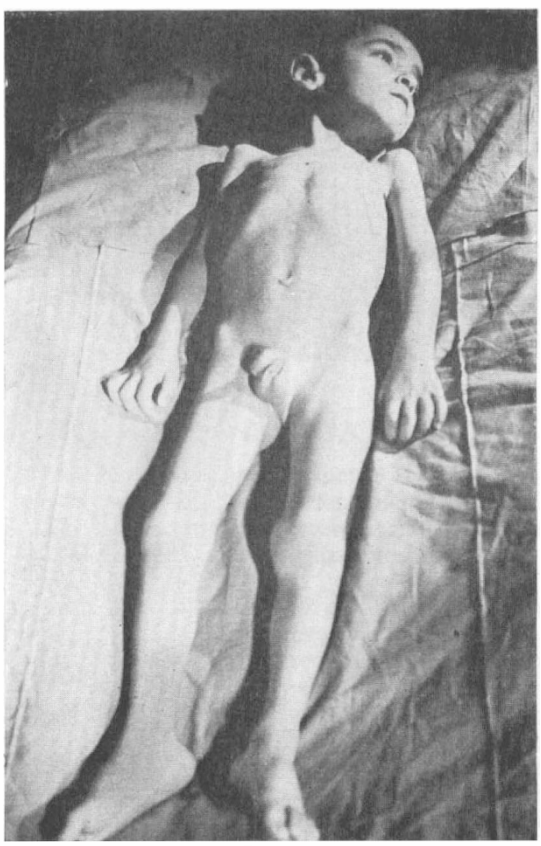

Fig. 1

A survey of literature by Karlen in 1962 revealed 27 anomalies of dens, of which three cases had absence of dens with permanent signs of cord damage and tetraplegia. It is interesting to note that trauma brought attention to all these cases (Roberts, 1933; Miyakawa, 1952). These patients were all between 8 and 58 years of age. We found a case of complete absence of dens in a boy aged 6 with tetraplegia (fig. 1). He made a full recovery following occipito-cervical fusion (fig. 4). Review of literature failed to reveal a similar report.

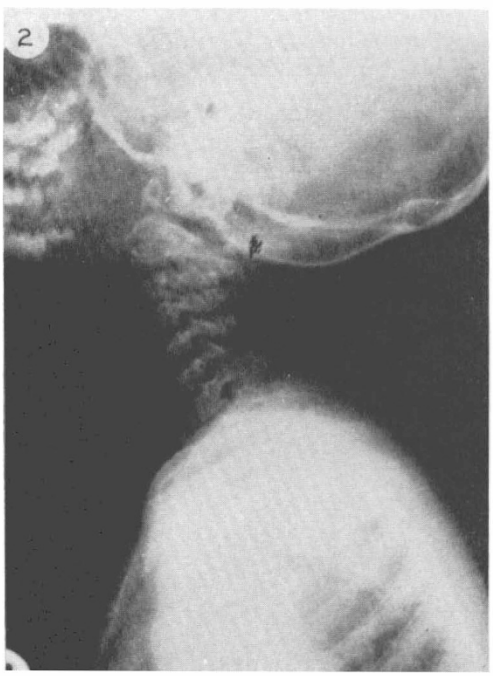

FIG. 2

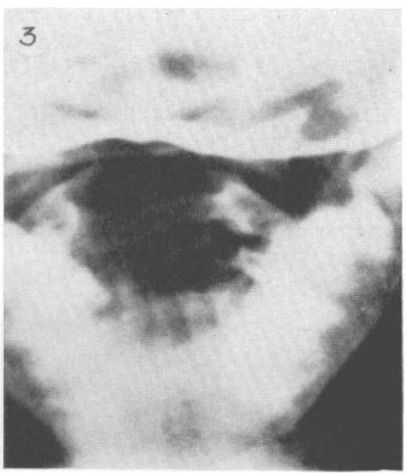

FIG. 3

CASE REPORT

R. N., a 6-year-old male, was brought to our clinic because of paralysis of both upper and lower limbs. There was no familial relevant history, but he was immature from birth. His birth weight was 2200 g. There was no apparent abnormality. His general condition gradually improved. He soon gained weight but even so, by the age of 6 never sat, stood or walked.

On examination, he was found to be a small boy, light weight with normal I.Q. He answered all questions clearly and complained of pain in the back of his neck. He could eat and drink comfortably. His respiration was normal in lying position. His upper and lower limbs showed marked spasticity and he had hyperreflexia in all four extremities. Bilateral Babinski and clonus were present. His head was normal. The neck was fixed, extended and turned to the left due to right torticollis. Movement of neck was impossible due to pain. He was unable to move either his hands or his legs.

Whenever he was held in a sitting position his neck dropped forward. In this position respiratory difficulties as well as aggravating spastic tetraplegia were produced. All respiratory troubles were relieved by putting him in a flat position. There was no sensory disturbance but there was partial incontinence of urine and faeces.

On investigation, routine blood tests were normal, skull and chest X-rays showed no abnormalities but X-ray of the cervical spine revealed complete absence of the dens (figs. 2 and 3). This was confirmed by tomogram. Myelography of the cervical spine did not show other abnormalities. Routine tests of the C.S.F. were normal.

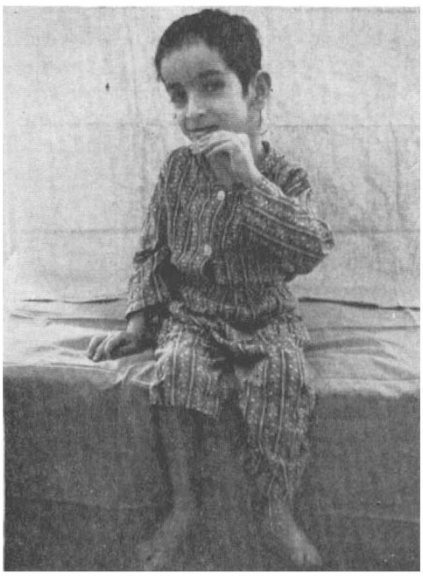

FIG. 4

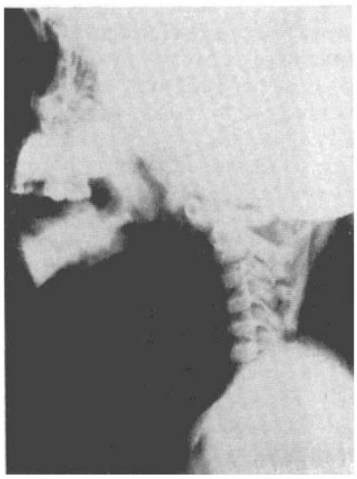

FIG. 5

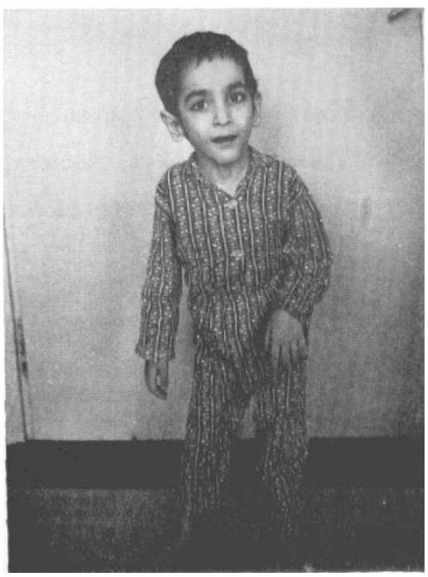

FIG. 6

Operative Treatment. Under general anaesthesia with endotracheal tube by posterior approach, first and second cervical vertebrae were exposed. There was complete instability of C1 on C2. Decorticating of posterior margin of occiput and C3, C4 and C5 were carried out. Two pieces of homograft bank bone approximately ($1 \times 0{\cdot}5 \times 5$ cm.) was fitted vertically between two slots in occiput and extended to C3-5. Some bone chips were also put in. The wound closed in layers without drain.

The patient was put in a posterior cast of half of the head and body with front fixation of forehead and chest, in order to provide secure immobilisation of the neck. Post-operative recovery was uneventful and all his symptoms gradually disappeared. The pain in his neck was relieved in less than a week. Spasticity of limbs completely disappeared in 8 weeks. Harmonic movements of extremities started in 3 months. Stability of the neck was satisfactory in 4 months (fig. 5). During this period the patient was able to sit and feed himself comfortably. In $5\frac{1}{2}$ months he was back to normal and walked when he was discharged from hospital (fig. 6).

DISCUSSION

Complete absence of the dens of the axis is an uncommon anomaly. The cases previously reported all came to light following relatively trivial neck injuries (Roberts, 1933; Ivie, 1956; Miyakawa, 1952; Sherk, 1967). Here we present a case in early childhood with severe torticollis and marked spastic tetraplegia. The clinical features may be misleading. It must be particularly differentiated from cerebral palsy and juvenile myopathy or tumours of the cervical cord.

SUMMARY

Embryological aetiology of anomaly of the dens of the axis discussed. The previous literature of this subject is briefly reviewed. A case of complete absence of dens of the axis with severe torticollis and spastic tetraplegia is presented. This 6-year-old patient recovered fully following occipito-cervical fusion.

RÉSUMÉ

L'étiologie des anomalies embryologiques de l'apophyse odontoïde est discutée. La littérature, sur ce sujet, est passée en revue.

Un cas d'absence complète d'apophyse avec torticolis sévère et tétraplégie spasmodique est présenté.

Ce malade, âgé de 6 ans, a récupéré complètement après une fusion occipito-cervicale.

ZUSAMMENFASSUNG

Die embriologische Ätiologie der Densanomal der Axis wird diskutiert und eine kurze Übersicht der einschlägigen Literatur wird gegeben. Ein Fall von komplettem Fehlen des Dens mit schwerem Torticollis und spastischer Tetraplegie wird beschrieben. Der 6 Jahre alte Patient wurde nach einer occipito-zervikalen Fusion völlig wiederhergestellt.

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