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Craniovertebral malformation with craniovertebral instability in an 8-month old baby

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An 8-month old female underwent cranial CT-scan for the evaluation of a severe occipital plagiocephaly secondary to congenital torticollis and was referred to us. The CT-scan confirmed that the plagiocephaly was positional but the luxation of both the C1 lateral masses was evident. The craniovertebral junction was studied by MRI and CT-scan and a severe malformation with craniovertebral instability was depicted. The atlas ossification nuclei had failed to join each other and C1 consisted of three separated parts: a schisis was present in the posterior arch with two laminae joining the lateral masses that were laterally and anteriorly displaced (left > right); the remnant of the anterior arch/dens apex consisted of a single anteriorly luxated bone fragment. On neurological examination, a mild tetraparesis was evident, and frequent episodes of sleep apnea were reported.
Conservative treatment was contemplated and an apposite neck brace was specifically manufactured, but it soon resulted not adequate to warrant the required stability.

Accordingly, craniovertebral fixation was decided: posterior arthrodesis was attempted by grafting autologous ribs that were fixed using silk threads on both sides from C0 to C2. It was decided that postoperative neck brace had to be maintained for at least 3 months. Postoperative CT scan documented the rift grafts were correctly placed, but 3 months later such grafts appears partially reabsorbed and 5 months postoperatively they completely disappeared. This was thought as consequence of the lightness of autologous ribs as well as of the inability to obtain absolute immobilization by the external neck brace.

Clinical conditions were unchanged. Reoperation was decided. This time, C0-C3 fixation was performed by placing two robust adequately modeled grafts of eterologous peroneal bone. Furthermore, now the patient was 14 month-old with fused cranial sutures, so that the postoperative immobilization could be achieved using a Halo vest jacket.

Six months later, the neuroimaging studies documented optimal craniovertebral arthrodesis. Eighteen months after the second surgery, the patient can freely walk without any orthoses. The torticollis completely resolved and the occipital plagiocephaly markedly improved.