Surgery versus watchful waiting in patients with craniofacial fibrous dysplasia: a meta-analysis
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CRD summary
The review concluded that optic nerve decompression surgery in patients asymptomatic craniofacial fibrous dysplasia (abnormal bone development of the skull) was associated with a worse visual prognosis compared with asymptomatic patients who were managed expectantly (monitored closely). Given uncertainties about the quality of the evidence base and the methods of analysis, caution is warranted when interpreting the authors’ conclusions.

Authors’ objectives
To compare the long-term outcome of patients with optic nerve compression due to craniofacial fibrous dysplasia who underwent surgery or were managed expectantly.

Searching
PubMed, CINAHL, Cochrane Central Register of Controlled Trials (CENTRAL), and Cochrane database of systematic reviews were searched from 1975 to 2010. Search terms were reported. Google and reference lists of retrieved articles were searched. The National Institutes of Health database records were also scanned. Articles in languages other than English that could not be translated were excluded.

Study selection
Eligible for inclusion were randomised controlled trials (RCTs), prospective cohorts, retrospective cohorts, case-control study designs, case reports and case series of patients with a histopathologic diagnosis of craniofacial fibrous dysplasia who underwent surgery or were managed expectantly. Studies had to have radiologically confirmed optic canal narrowing, pre- and post-treatment visual status (based on visual fields and visual acuity), and follow-up longer than four months.

The included studies considered patients with craniofacial fibrous dysplasia who underwent therapeutic surgery, prophylactic surgery or watchful waiting. The fibrous dysplasia was classified as being monostotic in 30% of cases and as polyostotic in 21% of cases. McCune Albright syndrome was diagnosed in 49% of patients. The median age at craniofacial fibrous dysplasia presentation was 20 years (range eight to 55 years).

Two reviewers independently performed the study selection.

Assessment of study quality
Two authors carried out selected aspects of quality assessment, but full details were not reported.

Data extraction
Data were extracted on blood pressure outcomes, and were used to calculate risk ratios (RRs) with 95% confidence intervals (CIs). Study authors were contacted for missing data.

Two reviewers independently performed data extraction, and disagreements were resolved by consensus.

Methods of synthesis
Meta-analysis was used to calculate pooled risk ratios and 95% confidence intervals. Clinical heterogeneity was assessed by comparison of effect sizes between symptomatic patients who underwent optic nerve decompression, asymptomatic patients who underwent optic nerve decompression, and asymptomatic patients who were managed expectantly. A narrative summary was also presented.

Results of the review
Twenty-seven published studies (119 patients) and a National Institute of Health study (122 patients) were included in the review. The duration of follow-up ranged from six to 228 months.
Of the clinically intact nerves, 85% were followed expectantly and 15% were surgically decompressed.

Improvement in visual function was reported in 67.4% of the clinically impaired nerves after surgery.

In patients who had normal vision, long-term stable vision was achieved in 75.6% of the operated nerves compared with 95.1% of the non-operated nerves (p=0.0003).

All clinically impaired optic nerves underwent therapeutic decompression.

Surgery in asymptomatic patients was associated with visual deterioration (RR 4.89, 95% CI 2.26 to 10.59).

Type of craniofacial fibrous dysplasia or growth hormone excess was not associated with postoperative worsening of visual outcomes in any of the treatment groups.

Results were confirmed by the NIH study.

Authors’ conclusions
The authors appeared to conclude that optic nerve decompression surgery in asymptomatic craniofacial fibrous dysplasia patients was associated with a worse visual prognosis compared with that of asymptomatic patients who were managed expectantly. They recommended that surgery be reserved for symptomatic patients.

CRD commentary
Inclusion criteria for the review were clearly defined. Several relevant data sources were searched. There was the potential for language bias, as only English language articles were included. Publication bias did not appear to have been assessed and could not be ruled out. Attempts were made to reduce reviewer error and bias throughout the review process.

Results were not presented for the quality assessment, which made it difficult to judge the reliability of the included studies. All of the included studies had small sample sizes. The evidence was mostly synthesised narratively. Although some form of meta-analysis was undertaken, full details were not presented; this made assessment of the robustness of the analysis difficult.

Given the uncertainties about the quality of the evidence base and the methods of analysis, caution is warranted when interpreting the authors’ conclusions.

Implications of the review for practice and research
Practice: The authors stated that expectant management was recommended in asymptomatic patients even in the presence of radiological evidence of optic nerve compression. Surgical decompression of the optic nerve should be reserved for symptomatic patients.

Research: The authors did not state any implications for research.

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This is a critical abstract of a systematic review that meets the criteria for inclusion on DARE. Each critical abstract contains a brief summary of the review methods, results and conclusions followed by a detailed critical assessment on the reliability of the review and the conclusions drawn.