



Melorheostosis

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Surgical Management of Melorheostosis: General Information and Considerations

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This document was prepared as general guidelines to educate patients and doctors about known surgical issues associated with melorheostosis. It is not intended as a recommendation of any particular treatment or surgery, nor as a substitute for evaluation between physicians and patients. Every melorheostosis patient is unique.

- Surgery to alleviate mechanical effects from melorheostosis in adults seems to be fairly effective. This is particularly true when the mechanical effects are mostly due to asymmetric bone growth, producing a deformity that can be corrected with bone procedures alone. As soft tissue causes of deformity increase, the need for osseous, soft tissue, and skin correction increases. In this case, surgery is less likely to produce satisfactory results, unless amputation is deemed an appropriate procedure.
- Surgery for the sole purpose of relieving pain (non-mechanical) in melorheostosis is rarely effective, unless that pain is a direct result of nerve pressure or irritation.
- Healing of osteotomies in melorheostotic bone is unpredictable and can be problematic. Bone stimulators or BMP products do not seem to help. Nevertheless, the most commonly used procedures for correction of melorheostotic deformity involve corrective osteotomies of bone.
- Contracture releases are more effective in adults than in children, and the results seem to improve with the use of rotation flaps, where possible. Pinning the involved joints may help, but does not assure that the contractures will not recur. The use of external fixators spanning the contracture area to provide corrective stretching may be preferable as a preliminary to surgery or even as sole treatment, but experience to date is very limited.
- Soft tissue releases alone in skeletally immature patients have a 100% "failure" rate in the literature. This statistic does not necessarily mean that they should not be done, but families need to be advised that the procedure may need to be repeated in the future. Soft tissue releases in the skeletally mature patient are not as affected by aggressive scar formation as is the case with children's tissues, but are best suited to release if the deformity is relatively recent.
- Melorheostosis has rarely been reported in the skull, face, ribs or spine.

If this information does not seem particularly encouraging, that is because the available information leads us to believe that surgical management for the orthopedic manifestations of melorheostosis is unpredictable and fraught with complications. This does not mean that surgery should never be undertaken, rather that the surgeon and the family must have a clear understanding of the risks and benefits involved and clear and realistic expectations about the proposed procedures.

The above observations are based on a retrospective review of 15 cases of surgery for the manifestations of melorheostosis in the upper extremities, review of the available English literature, a non-scientific review of 41 patient reported results of surgeries for melorheostosis from the www.melorheostosis.org website and the professional experience of the authors.

It is the authors' hope that this information will provide a basic framework of our current knowledge of this disease with regard to surgical management. Please consult your doctor to discuss the role of surgery in your specific situation.

