



FOP Australia is a small registered charity run by volunteers. We send concise updates twice yearly to any clinicians involved or interested in **Fibrodysplasia Ossificans Progressiva**

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Focus on: Montelukast and mast cells

Along with significant interest in macrophages, the role of mast cells in heterotopic ossification (HO) in FOP is an active area of research. The central role of inflammation in traumatic and FOP-related HO is recognised, but the role of mast cells is not well defined¹. Mast cell inhibition has been shown to result in a dramatic reduction of HO in a FOP mouse model^{2,3}. Further studies are required to identify the specific mediators/substances secreted by mast cells that exacerbate HO lesions¹. However these pre-clinical studies give rise to consideration of whether agents such as mast cell inhibitor cromolyn or leukotriene antagonist montelukast could help prolong mobility for patients with FOP. As explained in the current FOP treatment guidelines (available through a link on the top of every page of our website) the efficacy of these agents in reducing HO in FOP is not known. However the guidelines include some general information about use of these agents, along with contact details for international experts available to help you decide whether it is an appropriate option for your patient.

1. Levi B. *Sci Transl Med.* 2017 Sept 9(407) eaao6125. doi: 10.1126/scitranslmed.aao6125
2. Convente M et al. *e Miner Res.* 2018 Feb; 33(2):269-282. doi: 10.1002/jbmr.3304
3. Brennan T et al. *Bone.* 2018 April 109: 259-266. doi: 10.1016/j.bone.2017.08.023

Now on fopaustralia.org

The 'Living with FOP' section has information on a range of topics that your patient may find helpful, including guidebooks and resources, education, sport, transport and careers.

This information was compiled by FOP mother Julie Collins. We welcome any suggestions on further information to include to help our FOP families.

International Clinical Council on FOP

At the 2017 Drug Development Forum leading international FOP expert Prof Fred Kaplan from the University of Pennsylvania announced the formation of the International Clinical Council on FOP. This group includes expert clinicians from around the world, including Australia's Prof Matt Brown. The aims of the ICC include updating the current management guidelines. www.ifopa.org/international_clinical_council_on_fop

IFOPA Biobank development

Because biopsies and dental extractions trigger ossification in FOP, chances to access tissue samples for vital research are opportunistic and extremely rare. Transport delays from Australia and New Zealand to international research labs further limit possible contributions. FOP Australia and IFOPA are currently working on methods to maximise benefit from any possible samples from around the world. This includes development of an IFOPA Biobank. In the meantime, you have a patient who is likely to lose a tooth naturally, please contact us at info@fopaustralia.org and we will try to link you with researchers trialling transport strategies.

FOP Registry Update

In order to piece together vital knowledge about the natural history of FOP, thank you ensuring your patients update their data in the international FOP Connection Registry at fopregistry.org. Plans to launch the medical portal for clinician data are also underway. Learn more about this initiative in **Mantick N et al. Bone. 2018 April 109:285-290 doi: 10.1016/j.bone.2017.08.032**

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