Haemophilia - Management of Chronic Synovitis, Target Joints and Long-Term Surveillance of Musculoskeletal Health - Full Clinical Guideline

Reference no.: CG-HAEM/2023/010

1. Introduction

Prior to the availability of clotting factor concentrates, the natural history of arthropathy in severe haemophilia was well described, with recurrent bleeds into joints leading to progressive joint damage and ultimate destruction with associated functional problems. The prophylactic use of factor concentrates has dramatically modified the natural history of haemophilic arthropathy. The early use of prophylaxis can prevent joint bleeding and avoid the cycle of damage associated with recurrent haemarthrosis. Children who are treated on prophylaxis schedules often reach skeletal maturity with well-preserved joint function. However, in addition to the clinically obvious bleeds, recurrent subclinical episodes may also contribute to joint damage. Bleeding into joints leads to the development of synovitis, which is characterized by a painless chronic swelling of the affected joint. Such joints are more prone to recurrent bleeds and progressive joint damage.

2. Aim and Purpose

To prevent the development of and effectively treat chronic synovitis.

The term target joint is defined as a joint with recurrent bleeds. The ISTH definition suggests three or more spontaneous bleeds into a single joint within a consecutive six month period. The joint ceases to be a target joint when there have been less two bleeds into the joint within 12 consecutive months.

However in patients who are on prophylaxis any joint which has more than two bleeds over a six month period should be considered an ‘at risk joint’ requiring careful examination and investigation. A clotting factor trough level should be done. If prophylaxis is not already ongoing this should be started. If the patient is already on prophylaxis this should be intensified with more frequent or bigger doses of clotting factor. If there is no response to this a radioactive synovectomy (synoviorthesis) should be considered. ***This should be discussed with the CCC/regional haemophilia committee/NottinghamDerbyLincoln haemophilia MDT.***

The two commonest isotopes used are RE186 and Y90. They are short half lives of 2.7 to 3.7 days and a soft tissue penetration range of 1 to 5 mm. An infusion of clotting factor is necessary before the procedure. The isotope is introduced into the joint using a 16 or 18 gauge needle.

A reduction in the bleeding tendency is usually evident within two to three weeks of the procedure.

Long term surveillance of musculoskeletal health

Review by a physiotherapist is recommended as soon as a patient presents with or reports a joint bleed – advising and educating on the anticipated timeline for rehabilitation.

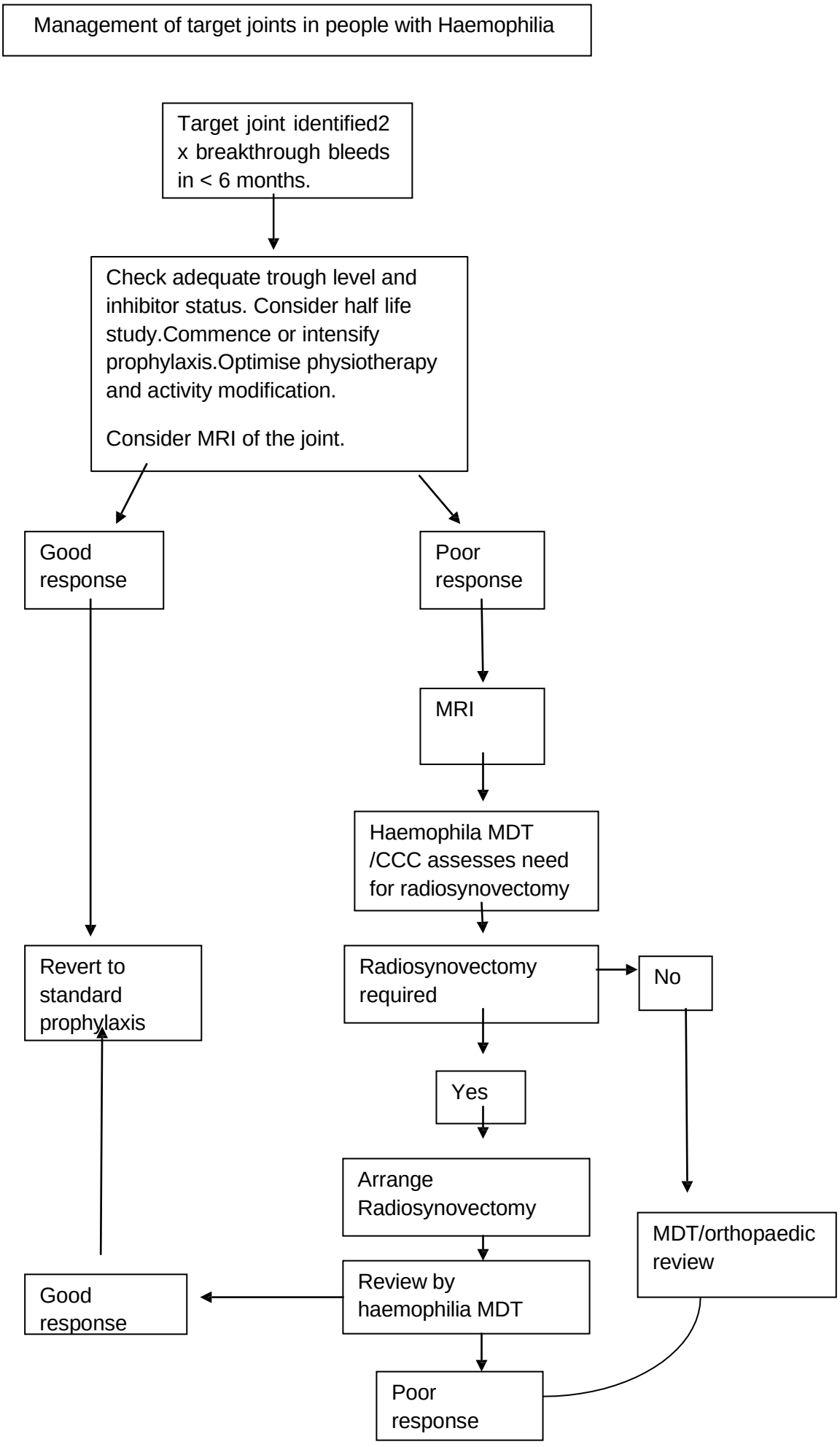
Initial assessment should be comprehensive – including history of trauma/injury as well as baseline joint health/function

Rest/immobilization should be monitored closely and kept to a clinical minimum

Cryotherapy to be used with the limb in elevation, preferably with crushed ice, and for no more than 20 mins per application. Gel packs and cyrocuffs can be used, but ice cubes or cooling blocks that do not mould to joint contour should be avoided.

Compression/elevation to be used according to patient comfort and monitored closely
Rehabilitation should focus on regaining dynamic joint control and towards improvement in baseline function. Techniques used in routine clinical practice can be used in persons with haemophilia. This should be preceded by a discussion with the haematologist to ensure appropriate clotting factor support if indicated.

All patients with severe and moderate bleeding disorder should have a yearly joint score performed by an experienced physiotherapist.


Management of target joints in people with Haemophilia
Target joint identified2 x breakthrough bleeds in < 6 months.
Check adequate trough level and inhibitor status. Consider half life study.Commence or intensify prophylaxis.Optimise physiotherapy and activity modification.
Consider MRI of the joint.
Good response
Poor response
MRI
Haemophila MDT /CCC assesses need for radiosynovectomy
Revert to standard prophylaxis
Radiosynovectomy required
No
Yes
Arrange Radiosynovectomy
MDT/orthopaedic review
Good response
Review by haemophilia MDT
Poor response

3. References

Guidelines for the management of acute joint bleeds and chronic synovitis in haemophilia A United Kingdom Haemophilia Centre Doctors' Organisation (UKHCDO) guideline. Haemophilia (2017), 1–10.

4. Documentation Controls

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