Management of Surgery and Dental Work in Patients with Inherited Bleeding Disorders

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Intended Audience

This document contains information and clinical guidelines for management of children attending the Oncology and Haematology department. It is to be used by all healthcare professionals within the Trust whenever they are caring for these children either in hospital or at home.

Purpose

To provide guidance, in according to national protocols, for the management of children with inherited bleeding disorders who require a surgical procedure including dental work.

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1. **Introduction**

Patients affected with inherited bleeding disorders (IBD) are at increased risk of bleeding with invasive procedures which includes minor and major surgery but also dental procedures and other invasive procedures which may not be considered surgical e.g. lumbar punctures. Bleeding can result in significant morbidity and carries a risk of mortality. Bleeding can largely be prevented or effectively managed by thoughtful forward planning. Good communication between surgical, anaesthetic and haematology teams is imperative to ensure the availability of specialist advice throughout the pre, peri and post-operative periods.

2. **Alerts**

All patients with IBD have an alert in their EDMS record which states that they are at increased risk of bleeding and surgical procedures must be discussed with haematology. The alert section includes an information record which details the diagnosis and any treatment notes e.g. usual factor concentrate, response to DDAVP, presence of inhibitors.

Patients and parents are counselled at the time of diagnosis and reminded at follow-up appointments regarding the need for surgical procedures and invasive dental work to be discussed in advance with the haematology team.

3. **Location of surgery**

Surgery in children with IBDs is scheduled at Sheffield Children’s Hospital where the specialist haematology team is based and there is access to laboratory assays and factor concentrates and other treatments to prevent and manage bleeding problems.

If surgery is suggested at another hospital in the region the clinician is asked to refer the child to a surgical team at SCH explaining the rationale for this advice.

Dental check-ups are permitted in the community but invasive procedures including extractions and those requiring an inferior nerve block should be referred to the Charles Clifford Dental Hospital (CCDH).

In general, patients will be admitted to the most appropriate surgical ward. However, if the patient is being admitted post-operatively purely for the monitoring and management of bleeding it may be more appropriate for the patient to be admitted to the haematology & oncology unit (Ward 6). The choice of ward for admission will be decided on a case by case basis by agreement between the haematology and surgery teams.

4. **Pre-operative planning**

All patients with an IBD who require surgery must be discussed with the haematology team. This is stated in the alert in the patient’s EDMS record as a prompt.

It is expected that the surgical team will contact the haemophilia team, preferably in writing, with as much notice as possible. Email correspondence is acceptable. It is important the patient is not listed for surgery until a plan has been made and the haemophilia team have indicated that the date is suitable for any haemostasis input required. In practice the surgical team may also ask the waiting list co-ordinator to check with the lead haemophilia consultant (currently Dr Payne) before confirming a date with the patient.

For urgent/emergency surgery a telephone request for advice should be made to the haematology SpR or consultant.
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Patients with haemophilia A or B requiring surgery or dental extractions need pre-operative inhibitor screening as per the inhibitor guideline with the result available prior to the day of surgery (Diagnosis and Management of Congenital Haemophilia with Inhibitors - H&O/11/1892 - CG1892). The timing of testing in relation to the surgery depends on the risk of developing an inhibitor e.g. a negative inhibitor screen within a couple of months of surgery may be acceptable for previously treated patients (PTPs) with >150 exposure days (ED) having surgery at a non-critical site but for a severe patient with < 20 EDs the sample should be taken between the last exposure and the surgery and in any case within a week of the date for surgery.

A plan for haemostatic aspects of surgery will be finalised by a consultant haematologist. For elective surgery the advice will be documented in a letter or email and recorded on EDMS. For patients with severe bleeding disorders or where treatment will be required over several days a formal plan is made using a proforma⁴. The plan will be sent to the surgical consultant and, if known, to the anaesthetist responsible for the list. If the anaesthetist has not been allocated at the time of writing the plan it is the responsibility of the surgical consultant to alert the anaesthetist to the plan and to disseminate to other relevant staff within their team e.g. specialist nurse, trainees. The haemostasis plan will state who to contact if unexpected bleeding occurs including out of hours.

The scheduling of a date and timing for surgery will need to take into account any need for specialist input on the day and the availability of relevant staff as well as any requirement for laboratory assays. Elective surgery in inhibitor patients who are at high risk of bleeding will only be scheduled for when a consultant haematologist is on site.

Blood product exposure will be avoided wherever there is a suitable alternative. If the use of blood products is expected then vaccination with hep A and B is recommended with a dose prior to the surgery date if timing allows.

5. Management of peri-operative haemostasis

Haemostatic management will be in line with national guidance where this is available¹-³. Additional information on the management of individual disorders can be found in Management of Haemophilia and Other Inherited Bleeding Disorders – H&O/11/1871 - CG1871

For patients >2yrs old with von Willebrand’s Disease (VWD) the use of DDAVP is preferred over factor concentrate provided that adequate levels can be sustained for the duration required.

For patients with mild or moderate haemophilia A, DDAVP will be used, where possible, to reduce the risk of an inhibitor following exposure to FVIII.

There is a risk of fluid retention and hyponatraemia following a dose of DDAVP. Fluid restriction is therefore required in the 24hrs following a dose of DDAVP to minimise the risk of hyponatraemia and fits.

The following is suggested as a guide to fluid restriction which MUST include oral and intravenous fluids

<table>
<thead>
<tr>
<th>Age</th>
<th>Maximal fluid intake /24hrs</th>
</tr>
</thead>
<tbody>
<tr>
<td>2-4yrs</td>
<td>&lt;750mls</td>
</tr>
<tr>
<td>5-10yrs</td>
<td>&lt;1 litre</td>
</tr>
<tr>
<td>&gt;10yrs</td>
<td>&lt;1.5litres</td>
</tr>
</tbody>
</table>
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If fluid restriction is not possible for the surgical procedure and post-operative course anticipated then DDAVP should be avoided and factor concentrate will be required.

If more than one dose of DDAVP is planned U&Es must be checked prior to each subsequent dose to check for hyponatraemia. If hyponatraemia occurs further doses of DDAVP must NOT be given.

Tranexamic acid is commonly used as an adjunct to reduce blood loss depending on the type of surgery. It can be administered with all forms of factor concentrate.

The management of surgery in patients with inhibitors is covered in the guideline Diagnosis and Management of Congenital Haemophilia with Inhibitors - H&O/11/1892 - CG1892

a) Dental Procedures

Invasive dental work such as extractions and procedures that require an inferior nerve block require haemostatic cover to raise factor levels to a target of 0.5iu/mL for haemophilia A, B and VWD. The administration of factor concentrate or DDAVP should be timed to aim for a peak level at the time of the procedure.

Oral tranexamic acid (TXA) may be used as a sole haemostatic agent (e.g in FXI deficiency) or as an adjunct to DDAVP or factor concentrate. Ideally, TXA should be started prior to the procedure with a dose given 2 hours pre-op and continued for 7-10 days post-procedure. For dental extractions under GA, an intravenous dose can be given at the time of induction of anaesthesia if necessary.

For older children who are able to use a mouthwash, TXA mouthwash (10 ml of a 5% solution) should be commenced just before the dental procedure to increase salivary levels, and continued 6-hourly for 7-10 days. The mouthwash should be swilled and expelled. The use of oral tranexamic acid (as above) to be swallowed is recommended in addition to the mouthwash in this older patient group.

b) Target levels for surgery

The following guidance applies for patients with haemophilia A and B and VWD. For patients with rare bleeding disorders refer to national guidelines where available and to individual patient plans.

The target level quoted is the trough factor level that should be maintained during the time periods specified i.e aim for the level not to fall below this value between doses. As levels will fall following a dose the half-life of the product used will need to be considered when calculating the dose and frequency of treatment

### Major surgery

<table>
<thead>
<tr>
<th>Stage</th>
<th>Target level iu/mL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-op</td>
<td>0.8-1.0</td>
</tr>
<tr>
<td>Days 1-3</td>
<td>0.6-0.8</td>
</tr>
<tr>
<td>Days 4-6</td>
<td>0.4-0.6</td>
</tr>
<tr>
<td>Days 7-14 (depends on surgical procedure)</td>
<td>0.3-0.5</td>
</tr>
</tbody>
</table>

### Minor surgery

<table>
<thead>
<tr>
<th>Stage</th>
<th>Target level iu/mL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-op</td>
<td>0.5-0.8</td>
</tr>
<tr>
<td>Days 1-5 (depends on surgical procedure)</td>
<td>0.3-0.8</td>
</tr>
</tbody>
</table>
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c) Lumbar punctures

Factor levels should be raised to near 1.00 iu/mL and this must be confirmed before the procedure – DO NOT proceed to LP if level is < 0.80 iu/mL. Thereafter the level should be maintained above 0.50 iu/mL for 24 hours.

6. References


4. Surgical Treatment plan H:\INFOAREA\haematology\CLINIC LETTERS\Haematology Patient Information\Haemostasis Thrombosis\General Haemophilia Information\Templates\7. Related Haematology & Oncology Unit Guidelines

Management of Haemophilia and Other Inherited Bleeding Disorders – H&O/11/1871 - CG1871
Diagnosis and Management of Congenital Haemophilia with Inhibitors - H&O/11/1892 - CG1892