Ashford and St. Peter's Hospitals
NHS Foundation Trust

Management of acute complications of sickle cell disease in children

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Guideline History		
Date	**Comments**	**Approved By**
Nov 2014	New guideline	Paediatric guideline group
April 2018	Complete review	Paediatric guideline group
June 2023	Changes made to format and content based on St George's Hospital Clinical Guideline for Children and Young People with Sickle Cell Disease	Paediatric guideline group
May 2025	Update regarding oxygen saturations – only provide oxygen if saturations <95%	Dr Claire Mitchell

Sickle Cell Disorders

Sickle cell disease (SCD) is now the most common serious genetic disorder in England, affecting over 1 in 2000 live births
The sickle cell disorders are a heterogeneous group of disorders affecting patients in whom haemoglobin S is the major haemoglobin. They include haemoglobin SS, SC, and S beta thalassemia.

It is important to make the diagnosis early to prevent and treat common complications effectively; some of them can be life threatening.

- Infection
- Acute Chest syndrome
- Painful Vaso-occulsive Crisis
- Splenic Sequestration
- “Aplastic” Crisis
- Stroke

Early vaccination and Penicillin prophylaxis significantly reduces the infection-related morbidity and mortality. Daily folic acid supplements help to minimise anaemia. Recently introduced Hydroxycarbamide therapy in minimises the frequency and severity of crises and the need for hospital admission.

The mainstay of management for sickle cell crisis are (see below for more information):

1. Fluid replacement
2. Pain relief
3. Antibiotics

*Sickle cell disorder patients are shared care with Tertiary centre (SGH).It is important to liaise with Dr Bhatti/Dr Mitchell (named consultants for Haemoglobinopathy) and Tertiary centre for advice.
Patients with Sickle cell disorder will have Passport status to the Paediatric ward.

Fluid Replacement:

Many patients with sickle cell disorders have reduced renal tubular concentrating ability. Continued fluid loss without adequate replacement causes a reduction in plasma volume with an increased blood viscosity and aggravation of sickling.

Minimum Requirements:
100 ml/Kg for the first 10 Kg body weight
50 ml/Kg for the next 10kg body weight
20ml/kg thereafter.
Fluids may be given orally or intravenously.
Use standard (5% dextrose 0.9% saline) re-hydration solution. Review the need for added potassium.

Maximum as per normal maintenance fluids calculation
Female: 80ml/hr
Male: 100ml/hr

• Children with sickle cell disease need individualised fluid regimes. They are often dry and will need additional fluids; conversely overzealous fluid replacement may make the situation worse by precipitating cardiac failure.

• The oral route should be used whenever possible but children with severe pain who are not settling, or who have abdominal symptoms should receive intravenous rehydration.

• Intravenous therapy should be stopped once the patient is stable and pain is controlled.

• Adequate oral intake should be documented.

Pain Management:

Parents often estimate their child's level of pain quite accurately using knowledge of their child's normal behaviour and responses. Parent's estimation of their child's pain is an important factor to be considered when managing the pain and planning the next stages.

• Healthcare Professionals can underestimate pain levels therefore assessment should be completed using a validated pain assessment tool taking into consideration the child's age and cognitive abilities.

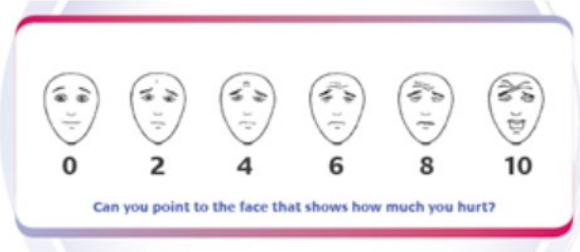
• This guideline is designed to guide practice for children and young people experiencing all levels of pain from mild to severe and also considers whether the child is treated at home or admitted to hospital. Individual assessment is therefore essential if quality care is to be provided.

An age appropriate pain scoring tool should be used.

The tools used are:

Visual Analogue Scale/Numerical rating scale – children >5 years

The Faces Pain Scale Revised – children above 3 years

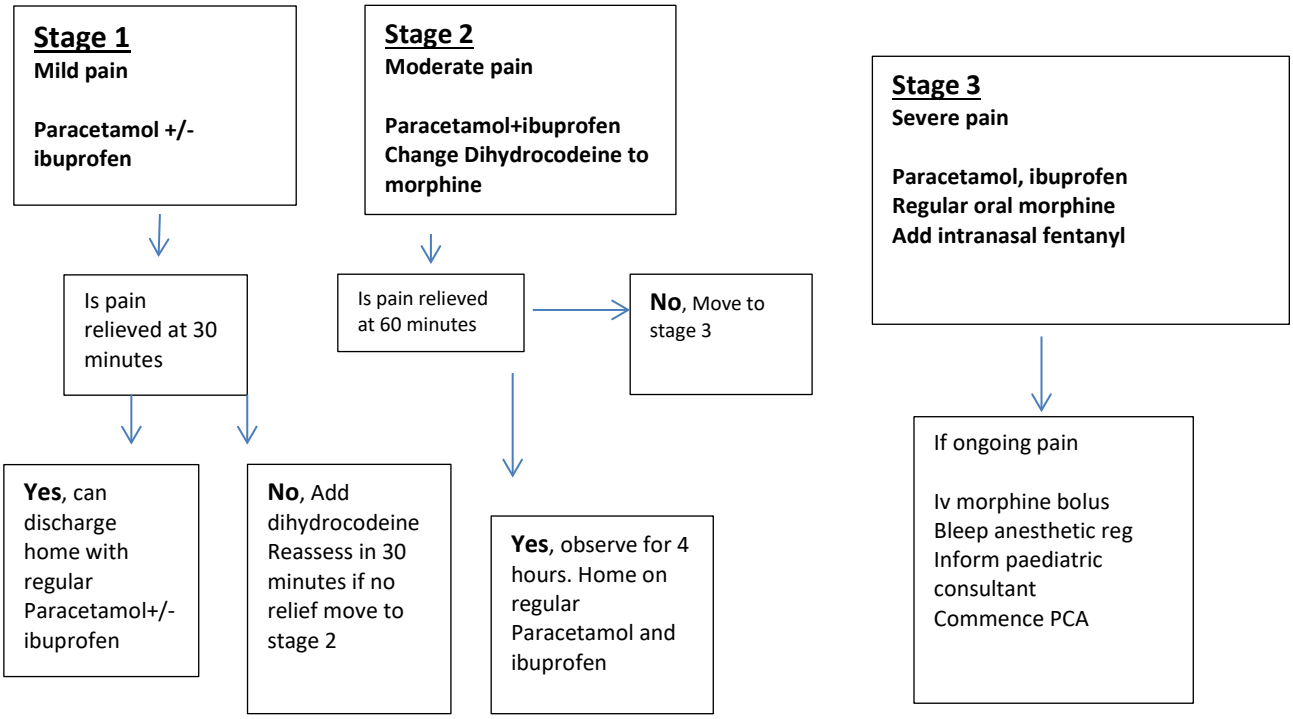


FLACC – for preverbal children

The Revised FLACC – for nonverbal children

Analgesia should be administered within 30 minutes of admission and aim for pain control within 60 minutes. Patients may have tried oral analgesia at home. A full assessment is needed to ascertain what has already been given and when.

Pain Management Flowchart



STAGE 1: MILD PAIN (Pain Score 1-3)

a) Paracetamol 15mg/kg QDS
b) Ibuprofen 10mg/kg TDS
+/-Dihydrocodeine 0.5mg/kg QDS

c) Non-drug methods can help a child or young person to relax and place the focus of their attention on something other than their pain. (NB. these methods should complement, NOT replace pain medicines). Methods where the child/young person plays an active rather than passive role can be more effective, e.g. the child reads the story not the nurse or parent.

Physical - stretching, application of heat pads (do not use cold packs as this can exacerbate sickling), massage, stroking

Behavioural - distraction (including play, blowing bubbles, and reading, music, and computer games), guided imagery often combined with relaxation

Information - Talking to children (think about their age, cognitive level and "What needs to be explained and talked about"?)

d) Reassess pain after 30 minutes - if not improved move onto next stage.
e) If assessed as clinically well with no other symptoms and tolerating adequate amount of oral fluids, the child can be discharged home on regular Paracetamol
f) Fluids – ensure full maintenance fluids are taken and tolerated.

STAGE 2: MODERATE PAIN (score 4-6)

a) Add oramorph 200mcg/kg 4 hrly; to be given 1 hour after Dihydrocodeine
b) Consider admission and baseline investigations
c) Maintain oxygen saturation above 95%
d) Encourage oral fluids or consider intravenous fluids
e) Check vital signs hourly for 6 hours and then 4 hourly if stable while on strong opioids

STAGE 3: SEVERE PAIN (pain score 7-10)

a) Consider intranasal fentanyl
b) Commence PCA/NCA Morphine. If PCA/NCA commencement is delayed – administer IV boluses of morphine 100mcg/kg
* Currently only PCA bolus without background is available on Ash ward, no NCA.
c) Continue nondrug methods and regular Paracetamol and NSAIDS
d) For details on drug doses and administration, refer to local paediatric pain management guidelines

Patient controlled Analgesia

Contact the inpatient pain service team in hours to help to set up PCA
Contact anaesthetic registrar covering maternity on bleep 5011 for out of hours
Child should be over 5 years and have the capacity to understand PCA.

Loading dose:

IV Morphine 0.1 mg / kg IV stat.
If not relieved in 15 minutes then second loading dose of 0.05 mg / kg. (May have up to 4 doses)

Bolus Dose:

This is the dose administered as a single bolus when the demand button is pressed by the patient.
Prescribe 1 mg/kg Morphine in 50 mls 0.9% Normal Saline (= 20 mcg/kg/ml) with lock out time of 5 minutes with 4 hourly limit.
For children over 50kg: Maximum dose 50 mg morphine in 50mls of 0.9% Normal Saline.

Background infusion - Currently not used in our paediatric department

Ensure no other opioid analgesia is administered with PCA UNLESS agreed by Lead Consultant for Inpatient Pain Service and Consultant paediatrician.
Ensure Paracetamol and NSAIDS are prescribed regularly
Ensure PRN naloxone is prescribed.
See PCA policy for detailed guidelines

Antibiotics:

Collect samples including: FBC, CRP, blood cultures, mycoplasma titres, urine and swabs for bacteriological studies, and viral screen, prior to starting antibiotics as below. Also, consider a CXR. If there has been foreign travel, consider a malaria screen.

Temperature >38 or suspicion of sequestration or chest syndrome

- Review child
- If signs of sepsis or crisis start IV ceftriaxone
- If clinically well consider increasing prophylactic penicillin from BD to QDS

If signs of chest infection

- Add IV clarithromycin
- Also do mycoplasma titres, if it is positive a 10 day course is required
- If clinically well can switch to oral co-amoxiclav + azithromycin

If suspicion of abdominal sepsis

- IV Ceftriaxone + metronidazole
- When clinically well can switch to oral ciprofloxacin and metronidazole
- Ascending cholangitis, cholecystitis, gut ischaemia are associated with gram negative and Salmonella infections

If suspicion of bone or joint infection

- IV Ceftriaxone + IV flucloxacillin
- Step down when clinically well to oral ciprofloxacin + flucloxacillin

Prophylactic penicillin

- Patients who are clinically well and are afebrile or have only a low-grade temperature should have their prophylactic penicillin increased from BD to QDS for 5 days.

Refer to paediatric antimicrobial guidelines+/- discuss with microbiologist

EMERGENCY MANAGEMENT- Potentially life threatening conditions

1. Febrile illness

• Fever in children with SCD is also a medical emergency. Splenic dysfunction can start as early as 6 months and by 5 years of age, most of them have functional asplenia increasing the risk of sepsis and death.

• Life threatening immune hemolysis can occur so children need to be monitored for splenic sequestration and acute chest syndrome

• Those with salmonella/staphylococcus aureus sepsis should be evaluated for osteomyelitis

• Children should also be evaluated for pneumonia due to typical and atypical organisms, malaria, particularly in children returning from holidays in Africa, urinary tract infection, acute cholecystitis and parvovirus B19

• Cholecystitis and gut ischaemia are associated with gram negative and salmonella infections. Iron overload can predispose to Yersinia and klebsiella infections

• Diagnostic problems can occur. It is common for a child with a simple acute painful episode to present with a fever and no obvious evidence of infection. Some young children present with painful swollen joints or areas of swelling in a long bone. In these cases, it may be difficult to differentiate between acute bone infarction due to sickling and osteomyelitis or septic arthritis.

2. **Acute Chest Syndrome** (Sequestration within the lungs)

Acute chest syndrome is an acute illness with

- **fever and/or respiratory symptoms**
- **PLUS**
- **New pulmonary infiltrate on CXR**

in a child with sickle cell disease.

This a life threatening condition, it is a common cause of death, and can be a postoperative complication in older children.
Patients can present with pain, cough, SOB, wheeze, fever, tachycardia, tachypnoea, and signs of lung consolidation.

Children with SCD frequently develop simple pneumonia. Generally, these children have unilateral chest signs, are only mildly unwell, have normal SaO_2 and have no chest pain. Over sedation with opiate use can result in hypoventilation, atelectasis and worsening hypoxia precipitating a chest crisis.

Investigations:
• FBC, Reticulocytes, and HbS %
• Biochemical profile
• Chest X-ray
• Sputum and blood cultures
• Serum for atypical serology

Management:
• Most important is prompt recognition of acute chest syndrome
• IV fluids as in painful crisis- but watch carefully for fluid overload continue with pain management, avoiding hypoventilation from over sedation.
• Maintain adequate oxygenation – start oxygen if sats <95% and aim to maintain >98%: monitor with pulse oximeter, regular bronchodilators by nebuliser (if wheeze present), chest physiotherapy and incentive spirometry.
• Treat underlying infection: Ceftriaxone at the higher dose plus IV azithromycin.
• Top-up transfusion, as discussed below.

3. Acute Splenic Sequestration (common in children under 3).

Characterised by sudden onset of abdominal pain, distension, sudden collapse, rapidly enlarging spleen and shock. May be precipitated by fever, dehydration, and hypoxia. Rapid sequestration of red cells can lead to sudden fall of the haemoglobin and death from hypoxic cardiac failure with pulmonary oedema. May have a more insidious onset.

Investigation:
- FBC
- reticulocytes
- cross-match
- blood-culture
- U&E
- store serum for virology.

Management:
- Red cell transfusion without delay (if in extremis uncross matched O negative).
- Transfusion should aim for Hb 80g/L
- Assess the need for volume expansion. Crystalloid should be used with caution as this may exacerbate heart failure.
- Broad-spectrum antibiotics to cover Pneumococcus and Haemophilus

NB) Hepatic Sequestration

Less common than splenic sequestration in children, but treated in the same way as acute splenic sequestration.

4. Aplastic Crisis

Onset of profound anaemia over 1 – 3 days without sequestration, due to transient parvovirus – induced marrow hypoplasia. Check Hb, retics, parvovirus DNA and antibody titres. If there is no reticulocyte response, transfuse. Immunity appears to be lifelong.

It is important to measure the reticulocyte counts when the child is in crisis to distinguish between aplastic crisis (low reticulocyte count), and sequestration (high reticulocyte count).

5. Acute neurological complications

All children with sickle cell anaemia should have transcranial Doppler studies from 2 years. Children can present with TIA, silent cerebral infarcts, subarachnoid bleed, intracranial bleed or acute ischemic stroke. They may present with severe headache, seizures, behavioural changes or focal neurological deficits.

Initial management in ED

Aim for stabilisation and transfer

- Manage airway, breathing, maintain saturation above 95%, IV access
- Regular monitoring of conscious level by GCS or AVPU and neurological status by PedNIHSS assessment tool
- Urgent blood samples – FBC, reticulocyte count, haematocrit, blood film, coagulation profile, blood group and cross match, blood glucose, blood gas, CRP, urea and electrolytes, LFT, bone profile, magnesium, LDH, blood culture, blood film, viral serology
- IVF 2/3 maintenance to avoid fluid overload
- CT scan within 1 hour
- Meningitis – consider LP and broad spectrum antibiotics+/- acyclovir
- Provide top up transfusion to Hb100g/L to improve oxygenation if exchange is likely to be delayed by >6 hours or if stroke occurs in the context of acute anaemia
- Urgent discussion with paediatric neurology and tertiary team
- Subarachnoid bleed, intracranial bleed – urgent neurosurgical opinion

6. Priapism

Priapism is painful erection of the penis lasting >1 hour. Prolonged priapism lasts for >3 hours, and stuttering priapism is repeated painful erections lasting >30 mins. This can start at night related to a full bladder. It can cause cumulative damage and may result in impotence.

Management

- **Home management**
  - Emptying the bladder, exercise and a warm shower or bath
  - If lasts >2 hours, severe pain or difficulty passing urine needs to seek urgent medical attention
- **ED management**
  - IV hydration + analgesia
  - Oral pseudoephedrine
    - 6-12 years 30mg 6 hourly
    - 12-18 year 60mg 6 hourly
  - Immediate referral to Surgical team
  - Catheterisation if unable to pass urine
- **Urological management**
  - If does not terminate, penile aspiration + irrigation with adrenaline within 4 hours of presentation
  - If continues needs Urology Consultant input
- **Management of stuttering priapism**
  - Increased fluids and frequent bladder emptying
  - Oral analgesia
  - Short course of pseudoephedrine can be used short term, seek urology advice

- **Discharge criteria**
  - Able to urinate
  - Penis more flaccid
  - Pain controllable on oral medications
  - Should be discharged home with 3-5 day course of pseudoephedrine
- **Follow up**
  - Paediatric urology clinic
  - Education
  - Home pseudoephedrine
  - If recurrent treatment considerations
    - Etilefrine 0.25mg/kg/dose BD orally
    - Leuprolein Acetate GnRH analogue 3.75mg IM every 4 weeks
    - Disease modifying therapy for sickle cell disease

7. Acute haematuria

Macroscopic haematuria may be due to urinary infection or papillary necrosis. Passing of renal papillae may cause renal colic and ureteric blockage. Haematuria can also occur in patients with sickle cell trait.

Investigations

- MSU to exclude infection
- Renal USS
- Hydrated intravenous urography- discuss with Urology team

Transfusion in sickle cell disease

Options include:

1. Simple/top-up transfusion,
2. Exchange transfusion
3. Hypertransfusion regimens – used in chronic setting to reduce HbS %

Transfusion is not recommended in uncomplicated painful crises unless there is a substantial drop in Hb from baseline (e.g. >20g/L or Hb < 50g/L).

Acute splenic and hepatic sequestrations are characterised by an acute fall in Hb >20 g/L and reticulocytosis

Over-transfusion (to Hb > 80 g/L) should be avoided in sequestration crises because of the risk of hyperviscosity due to the re-entry of sequestered red cells into the circulation.

Simple transfusion:

This is indicated for patients with mild or moderate chest syndrome and aplastic crisis aiming for a Hb level of no more than 100g/L. Use Kell compatible, rhesus compatible blood matched for antibody status.

Packed red blood cells cc = wt in kg x 3 x desired rise in Hb in g/dl.

Untoward Effects:

Hypertension convulsion syndrome if Hb is too high, especially in presence of high HbS%. Transfusion reactions.

Exchange transfusion:

If needed, as in severe chest crisis or stroke, the patient needs to be transferred to tertiary care hospital. The purpose of transfusion is to enhance oxygen-carrying capacity, improve tissue oxygen delivery, and reduce HbS concentration to reduce sickling and prevent progression to acute respiratory failure. Transfusion commonly results in impressive improvement within hours.

2. Supporting References

- NICE guidelines- Sickle cell disease: managing acute painful episodes in hospital
- https://starship.org.nz/guidelines/paediatric-pain-assessment
- St Georges Hospital- Clinical Guideline: Children and Young people with sickle cell disease who present to the Children's Emergency Department.
- www.rcpch.ac.uk/stroke-guideline
- https://www.nice.org.uk/guidance/cg143/evidence/full-guideline-pdf-186634333
- https://www.clinicalguidelines.scot.nhs.uk/nhsggc-guidelines/nhsggc-guidelines/haematologyoncology/sickle-cell-protocol/
- https://b-s-h.org.uk/guidelines/guidelines/management-of-acute-chest-syndrome-in-sickle-cell-disease

3. Supporting relevant trust guidelines

- Policy and Procedure for the Administration of Patient Controlled Analgesia (PCA) using the IVAC® PCAM® pump In the Paediatric Patient
- Management of Acute Pain in Children Best Practice Guidelines For Paediatric Inpatients
- Microguide

2. Guideline Governance

a. Scope

This guideline is relevant to all staff caring for all children from 0-18 years old across the emergency department, inpatient ward and outpatient department.

b. Purpose

i. This guidelines aims to facilitate a common approach to the management of children. At times deviation from the guideline may be necessary, this should be documented and is the responsibility of the attending consultant.

ii. This guideline is subject to regular review to ensure ongoing evidence based practice.

c. Duties and Responsibilities

All healthcare professionals responsible for the care of all children 0-18years should be aware of practice according to this guideline.

d. Approval and Ratification

This guideline will be approved and ratified by the Paediatric Guidelines Group.

e. Dissemination and Implementation

i. This guideline will be uploaded to the trust intranet 'Paediatric Guidelines' page and thus available for common use.

ii. This guideline will be shared as part of ongoing education within the Paediatric Department for both medical and nursing staff.

iii. All members of staff are invited to attend and give comments on the guideline as part of the ratification process.

f. Review and Revision Arrangements

a. This policy will be reviewed on a 3 yearly basis by the appropriate persons.

b. If new information comes to light prior to the review date, an earlier review will be prompted.

c. Amendments to the document shall be clearly marked on the document control sheet and the updated version uploaded to the intranet. Minor amendments will be ratified through the Paediatric Guidelines Group. A minor amendment would consist of no major change in process, and includes but is not limited to, amendments to documents within the appendices.

g. Equality Impact Assessment

Background • Who was involved in the Equality Impact Assessment
Author and the supervising consultants.
Methodology • A brief account of how the likely effects of the policy was assessed (to include race and ethnic origin, disability, gender, culture, religion or belief, sexual orientation, age) • The data sources and any other information used • The consultation that was carried out (who, why and how?)
All groups of staff and patients were taken into consideration and there is no bias towards or against any particular group.
Key Findings • Describe the results of the assessment • Identify if there is adverse or a potentially adverse impacts for any equalities groups
There is no evidence of discrimination.
Conclusion • Provide a summary of the overall conclusions
There is no evidence of discrimination.
Recommendations • State recommended changes to the proposed policy as a result of the impact assessment • Where it has not been possible to amend the policy, provide the detail of any actions that have been identified • Describe the plans for reviewing the assessment
This guideline is appropriate for use.

h. Document Checklist

To be completed (electronically) and attached to any document which guides practice when submitted to the appropriate committee for approval or ratification.

Title of the document:

- Management of Acute Complications of Sickle Cell Disease in Children

Policy (document) Author: Dr Rose Cherian, Dr Naomi Cobbold

Executive Director: N/A

		Yes/No/ Unsure/NA	Comments
1.	**Title**		
	Is the title clear and unambiguous?	**Yes**	
	Is it clear whether the document is a guideline, policy, protocol or standard?	**Yes**	
2.	**Scope/Purpose**		
	Is the target population clear and unambiguous?	**Yes**	
	Is the purpose of the document clear?	**Yes**	
	Are the intended outcomes described?		
	Are the statements clear and unambiguous?	**Yes**	
3.	**Development Process**		
	Is there evidence of engagement with stakeholders and users?	**Yes**	
	Who was engaged in a review of the document (list committees/ individuals)?		**Clinical Guidelines Meeting**
	Has the policy template been followed (i.e. is the format correct)?	**Yes**	
4.	**Evidence Base**		
	Is the type of evidence to support the document identified explicitly?	**Yes**	
	Are local/organisational supporting documents referenced?	**Yes**	
5.	**Approval**		
	Does the document identify which committee/group will approve/ratify it?	**Yes**	
	If appropriate, have the joint human resources/staff side committee (or equivalent) approved the document?		
6.	**Dissemination and Implementation**		
	Is there an outline/plan to identify how this will be done?	**Yes**	
	Does the plan include the necessary training/support to ensure compliance?		
7.	**Process for Monitoring Compliance**		
	Are there measurable standards or KPIs to support monitoring compliance of the document?		
8.	**Review Date**		

		Yes/No/ Unsure/NA	Comments
	Is the review date identified and is this acceptable?	**Yes**	**3 years**
9.	**Overall Responsibility for the Document**		
	Is it clear who will be responsible for coordinating the dissemination, implementation and review of the documentation?		
10.	**Equality Impact Assessment (EIA)**		
	Has a suitable EIA been completed?		

Committee Approval (Paediatric Guidelines Group)			
If the committee is happy to approve this document, please complete the section below, date it and return it to the Policy (document) Owner			
Name of Chair	**Dr Claire Mitchell**	**Date**	**09/12/2023**
Ratification by Management Executive (if appropriate)			
If the Management Executive is happy to ratify this document, please complete the date of ratification below and advise the Policy (document) Owner			
Date: n/a			