Bringing to life the b

t e Englis language T e ational ibra f or Healt and t e ational Guidelines Clearing House were also sear ed for relevant guidelines ere possible, re o endations are based on available eviden e, areas w ere eviden e is la ing are annotated as good pra ti e points (designated by ati .

## 4. Preconception care

*at are t e add t ona r sks to t e wo an and baby*

SCD is asso iated wit bot aternal and fetal o plications and is asso iated wit an in reased in iden e of perinatal ortalit , pre ature labour, fetal growt restri tion and a ute pain ul ises during pregnan y.

T e assess entf or ~ ron i~ dise ase ~o pli~ations s oul d in~lude_

- S~reening f or pul onaŷ y pertension wit e~ o~ardiograp y. T e in~iden~e of pul onaŷ y pertension is in~reased in patients wit SCD and is asso~iated wit in~reased ortaliŷ. ¶A tri~uspid regurgitant jet velo~iŷ of ore t an . /se~ond is asso~iated wit a, ig ris of pul onaŷ y pertension S~reening s oul d be pef or e d if it is as not been ~arried out in t e lastŷ ear
- Bloo d pressure and urinaŷ sis s oul d be pef or e d to identifŷ wo en wit y pertension and/or proteinuria, enal and liverf un~tion tests s oul d be pef or e d annuaŷ to identifŷ si~ le nep ropat y and/or deranged epati~f un~tion
- etinal s~reening proliferative retinopat y is ~o on in patients wit SCD, espe~iaŷ patients wit HbSC, and ~an lead to loss of vision T ere is no ran do ised eviden~e on w et er routine s~reening s oul d be pef or e d or if patients s oul d be s~reened onŷ if t e perien~e visuaŷ sy pto s, but we re~o end t at wo en are s~reened pre~on~eptuaŷ.
- S~reening f or iron overloa d In wo en w o aye been ultipŷ transf used in t e past or w o aye a, ig f erritin level T * ~ardia~ agneti~ reson an~e i aging ŷ be elpf ul to assess boŷ iron loading Aggressive iron ~ elation bef ore ~on~eption is advisable in wo en w o are signfi~antŷ iron loaded
- S~reening f or red ~ell antibodies ed ~ell antibodies ŷ indi~ate an in~reased ris of ae oŷ ti~ disease of t e newborn

### *at s t e portance of genet c screen ng and w at procedure s are nvo ved*

**Women and men with SCD should be encouraged to have the haemoglobinopathy status of their partner determined before they embark on pregnancy. If identified as an 'at risk couple', as per National Screening Committee guidance, they should receive counselling and advice about reproductive options.** **D**

Gener al pra~titioners aye a, eŷ role to plaŷ in partner s~reening and geneti~ ~ounselling o en s oul d be en~ourage d to aye t e ae oglobinopat y status of t eir partner teste d f a, partner is a, ~arrier of , or ff e~te dŷ , a, ajor ae oglobinopat y , t e ~ouple s oul d re~eive appropriate ~ounselling regarding t e ris of aying ff e~te d off spring (T able ). T e et ods and ris s of prenatal diagnosis and ter ination of pregnan y s oul d be dis~usse d wit t e ~ouple In addition, t eŷ s oul d re~eive ~ounselling about t e ayailabiliŷ of prei plantation geneti~ diagnosis and ref erre d f or t is f appropriate partners will not alwaŷ s be ayailable or willing to undergo pre~on~eptuaŷ testing o en wit SCD s oul d be aware t at if t eir partner's status is un nown, t ef etus s oul d be treate d as ig ris f or a, ae oglobinopat y. Sper donors s oul d also be s~reene d f or ae oglobinopat ies f or ~ouples ~onsidering in vitro f ertilisation

Furt er inf or ation ~an be obtaine d f ro t e HS Si~ le Cell &T alassae ia, S~reening progra, e website or t e progra, e's *Handbook for Laboratories*

**Table 1.** Conditions requiring counselling when the other is affected by SCD[35]

| Condition | |
|---|---|
| H . | C rn rst t np rtn r . . . r qu r s r \ rr \ or \ ouns \ n n o \ ro pr n t \ r . nos s |

pregnant while taking hydroxycarbamide, it should be stopped and a level ultrasound performed to look for structural abnormality, but termination is not indicated based on exposure to hydroxycarbamide alone.

It is recommended that women receive low-molecular-weight heparin during hospital admission

Non-steroidal anti-inflammatory drugs (NSAIDs) should be prescribed only between and weeks of gestation owing to concerns regarding adverse effects on fetal development

*Additional care should be provided during routine antenatal appointment*

**Antenatal appointments for women with SCD should provide routine antenatal care as well as care specifically for women with SCD.**

patient ^o ort, in^luding alloi unisation, · delay^ed transfusion rea^tions, trans ission of ife^tion and iron overloa^d A rando ise^d ^ontrolled trial and a retrospe^tive stud^ aye de onstrate^d t at prop^y la^ti^ transfusion de^rease^d t e in^iden^e of aternal pai^ful ^rises but did not if luen^e f etal or aternal out^o e A S^ste ati^ review^ indi^ate^d t at t ere is insuff i^ient evidence to draw ^on lusions about t e role of transfusion in pregnan^y.

Evidence level -

**Table 2.** Specific antenatal care for wo en with SCD

| Appointment | Care for women with SCD during pregnancy |
|---|---|
| . . ts. ou^r . pp n t<br>tt^ _rst ppo nt ^nt | . ^ r n.or ton rv^ nr support nr^ ton to opt sn ^ n r ^ t. _D |
| .r rl^ r or<br>.osp t ppo nt ^nt | . ^ rp rtn rt st n _not ^r r ron : r v^ p rtn rr sults _ v ^ nr s.uss. _D _<br>pprop t ID<br>. ^ n^ . stor to^ st. s^ ^t nt o_ CD nr ts^o p^ tons<br>^ v ^b^ tons nr ts^o p^ tons: t ^n . rro^ r ACE n. tors or A Bs<br>t^ s s. ou^r ^ lstopp r _D<br>. o ^ ns. ou^r ^ r r ^ t ^n r ^ o^ ^ r nr nt ot^ prop. ^ s _no^ ontr nr^ ton _D<br>D s.uss v ^ n tons _DI<br>. ^ rr tn^ nr orr n^ nr or^ nr ^ ss ss ^ nts _t^ s . v not^^ np r.or ^ r nt^<br>pr v ous ^ r _D<br>Do^ u ^ nt s^ In o. ^ ns tur tons nr. oor pr ssur<br>. ^ nr . ^ .or^ utur |
| - ^^ ^s | Con_r v . t n^ v o_t^ n^r s r^r s^o_ s. rr^ ^ _D |
| . . ts. ou^r . pp n t<br>t^ . oo^n pp nt ^nt | D s.uss n.or ton ^r u^ ton nr ^r v^ . out. o . CD ^^ ^ t pr^ n n^ _D |
| .^^ r ^ t^ . pr n^ n^ l<br>n. . . rs^q st tr^ s _poss. ^ | . ^ v^ l p rtn rr sults nr r s.uss. _D _ pprop t _D l<br>B s^ n r n^ un^ ton t st urn^ prot n^ r t n^ n r to^ v r un^ ton t st nr ^ rr t n s. ou^r. ^<br>p r.or ^ r _D ll<br>E. t nr r r r ll p. ^ not p _not pr v ous p r.or ^ r _D<br>Con_r t. t ^^ ^ tons ro _rst v st r ^ o p t _D<br>Cons^ n^ o r os sp r n_ro t ^^ ^s o_^ st ton _D |
| . l^^ ^s ls^ r ^ p us<br>ut r s. p n r r v | . out n sl p r_ICE r p t _k<br>ut r s. p n r r v _ onsu t nt q st tr^ n nr . ^ to q st |
| t l^^ ^s ls^ r ^ p us<br>ut r s. p n r t | D t ^ r ut r soun r sp r_ICE nt n t^ un^ n<br>. ^ p t . ^<br>. ^ p t FBC |
| t . ^^ ^s s^ ut r s. p n r<br>t | ^ tr soun r on tor n o_ t^ ro t. nr not^ ur vou ^ –<br>. ^ p t . ^ |
| t ^^ ^s s^ r ^ | . out n ^ . ^ ^ n^ ur n . oor pr ssur nr urn^ ss |
| t_ ^^ ^s s^ ut r s. p n r<br>t | ^ tr soun r on tor n o_ t^ ro t. nr not^ ur vou ^<br>. ^ p t . ^<br>. ^ p t FBC nr . roup nr nt or s. r n |
| ^^ ^s ls^ r ^ nr o_ r<br>nt n t^ ^ ss s | . out n ^ . ^ ^ n^ ur n . oor pr ssur nr urn^ ss |
| t ^^ ^s s^ ut r s. p n r<br>t | . lout n ^ . ^ ^<br>^ tr soun r on tor n o_ t^ ro t. nr not^ ur vou ^<br>. ^ p t . ^ nr FBC |
| . ^^ ^s s^ r ^ | . out n ^ . ^ ^ n^ ur n . oor pr ssur nr urn^ s |
| ^^ ^s s^ ut r s. p n r<br>t | . lout n ^ . ^ ^<br>^ tr soun r on tor n o_ t^ ro t. nr not^ ur vou ^<br>. ^ r n or ton nr r v^ . out<br>_ t ln . or^ nr n^ ^ nto_t^ . rt.<br>_ n^ ^ s nr n^ st^ s : rr n^ n^ st^ t^ ss ss ^ nt<br>_ ^ r o_ . _t r. rt. |
| _ ^^ ^s s^ r ^ nr<br>q st tr^ n | . out n ^ . ^ ^<br>. ^ o ^ nr nr u^ ton o_ . our or^ ^ s r ns^ ton ^ t ^^ n _ nr . ^^ ^s o_^ st ton |
| . ^^ ^s s^ r ^ | . out n ^ . ^ ^ nr r^ o ^ nr r^ ^ v r . ^^ ^s o_^ st ton |
| . ^^ ^s s^ q st tr^ n | . out n ^ . ^ ^ nr o_ r^ t^ on tor n _t^ o nr ^ n s r^ v r . ^^ ^s o_^ st ton |

ACE^ n ot^ ls n^ on v rt n^ n^ ^ : A B^ In ot^ ls n s nr^ r ptor^ o^ ^ le FBC^ u^ . oor^ ount _d t^ o n: . ^ ^ r str urn:
_ ICE^ _ ton Inst tut _or H t. nr O n^ E^ ^ n^ : . D pr n t^ r ^ noss. CD^ s^ ^ ^ r s s

'Top-up' transfusion is indicated for women with acute anaemia. Acute anaemia may be attributable to transient red cell aplasia, acute splenic sequestration or the increased rate of lysis and volume expansion encountered in SCD. There is no absolute level at which transfusion should be undertaken, and the decision must be made in conjunction with clinical findings, but the oglobin under 6g/dl or a fall of over 6g/dl from baseline is often used as a guide to transfusion requirement.

Exchange transfusion for ACS was demonstrated to be effective in one prospective randomised trial and is accepted as best practice.

Exchange transfusion is also indicated for acute stroke.

The decision to recommend transfusion should be made by an experienced haematologist and obstetrician. Indications for transfusion are summarised in Table.

Alloimmunisation (the formation of antibodies to red cell antigens), is common in SCD, occurring in

Fluid intake of at least 3 l/24 hours should be ensured. This can be taken either orally or intravenously if the woman is not able to take adequate oral fluids. There is a risk of fluid overload in women with pre-eclampsia, senior experienced staff should be involved in managing the fluid balance of these women. Oxygen saturations should be monitored and facial oxygen should be prescribed if oxygen saturation falls below the woman's baseline or below 96%. There should be early recourse to intensive care if satisfactory oxygen saturation cannot be maintained by facial or nasal prong oxygen administration.

The woman should be assessed for infection. Therapeutic antibiotics should be prescribed if the woman is febrile or there is a high clinical suspicion of infection. White blood cell counts are often raised in SCD and do not necessarily indicate infection. Thromboprophylaxis should be provided to women with SCD who are admitted to hospital with painful crises. Other adjuvants may be required to treat the adverse effects of opiates, such as antihistamines to treat itching or laxatives to prevent opiate-induced constipation, and antiemetics may be required. As the painful crisis resolves, most women are able to reduce their opiate requirement rapidly, but this should be guided by the woman's previous experience.

Opiates are not associated with teratogenicity or congenital deformation but may be associated with transient suppression of fetal movement and a reduced baseline variability of the fetal heart rate. Where a woman has received prolonged administration of opiates in late pregnancy, the neonate should be observed for signs of opioid withdrawal.

### *What are the other acute complications of SCD and how are they treated*

**All patients, carers, medical and nursing staff should be aware of the other acute complications of SCD, including ACS, acute stroke and acute anaemia.**

**Each hospital should have a protocol in place for the management of ACS in pregnancy, including the use of transfusion therapy.**

SCD is associated with other acute complications including ACS, stroke and acute anaemia. In the pregnant woman, these complications should be managed in the multidisciplinary setting by an obstetrician and anaesthetist, and guidance on the management of these complications can be found in the relevant RCOG standards.

After acute pain, ACS is the most common complication, reported in 25% of pregnancies with ACS is characterised by respiratory symptoms such as tachypnoea, chest pain, cough and shortness of breath in the presence of a new infiltrate on the chest X-ray. The signs and symptoms of ACS are the same as those of pneumonia, so both should be treated simultaneously. Acute severe infection with the H1N1 virus in pregnancy can cause a similar clinical picture, and investigation and treatment for this should be instituted.

A^ute stro e, bot itf ạr^tive ạn ḍ ạe orr ạgi^, is ạsso^iạte ḍ wit SCD

**The relevant multidisciplinary team (senior midwife in charge, senior obstetrician, anaesthetist and haematologist) should be informed as soon as labour is confirmed.**

**Women should be kept warm and given adequate fluid during labour.**

**Continuous intrapartum electronic fetal heart rate monitoring is recommended owing to the increased risk of fetal distress which may necessitate operative delivery.**

T ere are no rand o ised ^ontrolled trials wit regard to pla^e of birt f or wo en wit SCDT ere is an in^reased f requen^y of si^ le ^ell ^risis and ACS in t e intrapartu periodT ere is an in^reased ris of pain ful ^risis wit protra^ted labour ( ore t an ours, but t is is often se^ondary to de y dration In t is situation, if t e wo an is well y drated and labour is progressing, t e labour s ould be ^are fully supervised ^aesarean se^tion s ould be ^onsidered if labour is not progressing well and delivery is not i inent

During labour, if oral y dration is not tolerated or is inadequate, intravenous f luids s ould be ad inistered using a f luid balan^e ^ art to prevent f luid overload enous a^^ess ^an be diffi^ult, espe^ially if t e aye ad ultiple previous ad issions, and as su^ anaest eti^ review/intravenous a^^ess s ould be obtained earlyT e de and of or o y gen is in^reased during t e intrapartu period and t e use of pulse o i etry to dete^t y po ia, in t e ot er is appropriate during labour Arterial blood gas analysis s ould be perf or ed and o y gen t erapy institute if o y gen saturation is or less

routine antibioti^ prop y la is in labour is ^urrently not supported by eviden^e, but our y observations of vital signs s ould be perf or ed A raised te perature (over . C, requires investigationT e ^lini^ian s ould aye a low t res old to ^o en^e broad-spe^tru antibioti^s

T ere are no rand o ised ^ontrolled trials wit regard to interventions during labour f or wo en wit SCD E perien^e reported in ^o ort observational studies f ro si^ le ^ell ^entres in Ja, ai^a, t e SA and t e re^o end ^lose observation, as des^ribed above Continuous ele^troni^ f etal eart rate onitoring is re^o ended be^ause of t e in^reased rate of stillbirt , pla^ental abruption and ^o pro ised pla^ental reserve

### *at s t e opt a ode of ana ges a and anaest es a*

**Women with SCD should be offered anaesthetic assessment in the third trimester of pregnancy.**

**Avoid the use of pethidine, but other opiates can be used.**

**Regional analgesia is recommended for caesarean section.**

pregnant wo en wit SCD are at ris of end organ da, age as well as e perien^ing a, ig er rate of ^aesarean se^tion General anaest esia, ^arries additional ris s be^ond t e nor al obstetri^ ^ase and s ould be ayoided w ere possible regional anaest esia, during labour a, redu^e t e ne^essity of general anaest esia, f or delivery. It is also li ely to redu^e t e need f or ig doses of opioids if t e wo an as si^ le-related pain in t e lower bo y. An anaest eti^ assess ent in t e t ird tri ester is warranted pet idine s ould be ayoided

## 7. Postpartum care

*at s ou d be t e opt u care post de very*

**In pregnant women where the baby is at high risk of SCD (i.e. the partner is a carrier or affected), early testing for SCD should be offered. Capillary samples should be sent to laboratories where there is experience in the routine analysis of SCD in newborn samples. This will usually be at a regional centre.**

**Maintain maternal oxygen saturation above 94% and adequate hydration based on fluid balance until discharge.**

**Low-molecular-weight heparin should be administered while in hospital and 7 days post-discharge following vaginal delivery or for a period of 6 weeks following caesarean section.**

**The same level of care and vigilance should be maintained as has been described for antenatal care,**

. Pauling L, Itano HA, Singer SJ, Wells IC Sickle cell anaemia, a molecular disease *Science* , – .
. Stuart MJ, Nagel RL Sickle cell disease *Lancet*

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## APPENDIX

Clini^al guidelines are 'sy ste ati^ally developed state ents w i^ assist ^lini^ians and wo en in a, ing de^isions about appropriate treat ent for spe^ifi^ ^onditions'. Ea^ guideline is sy ste ati^ally developed using a, standardised et odology. E a^t details of t is pro^ess ^an be found in Clini^al Governan^e Adviˆe o _ *Development of RCOG Green-top Guidelines* (available on t e RCOG website at r p r o or ś nés). T ese re^o endations are not intended to di^tate an e ^lusive ^ourse of anage ent or treat ent T ey ust be evaluated wit referen^e to individual patient needs, resour^es and li itations unique to t e institution and variations in lo^al populations It is oped t at t is pro^ess of lo^al owners ip will elp to in^orporate t ese guidelines into routine pra,ti^e Attention is drawn to areas of ^lini^al un^ertainty w ere furt er resear^ ig t be indi^ated

T e eviden^e used in t is guideline was graded using t e s^ e e below and t e re^o endations for ulated in a, si ilar fas ion wit a, standardised grading s^ e e

| C . . . on o . . . n . | r o r o n . on |
|---|---|
| ++ Hig -quality eta,analy ses, sy ste ati^ reviews of rando ised ^ontrolled trials or rando ised ^ontrolled trials wit a, very low ris of bias | **A** At least one eta,analy sis, sy ste ati^ review or rando ised ^ontrolled trial rated as ++ and dire^tly appli^able to t e target population, or<br>A sy ste ati^ review of rando ised ^ontrolled trials or a, body of eviden^e ^onsisting prin^ipally of studies rated as + dire^tly appli^able to t e target population and de onstrating overall ^onsisten^y of results |
| + Well-^ondu^ted eta,analy ses, sy ste ati^ reviews of rando ised ^ontrolled trials or rando ised ^ontrolled trials wit a, low ris of bias | **B** A body of eviden^e in^luding studies rated as ++ dire^tly appli^able to t e target population, and de onstrating overall ^onsisten^y of results, or<br>E trapolated eviden^e fro studies rated as ++ or + |
| - Meta,analy ses, sy ste ati^ reviews of rando ised ^ontrolled trials or rando ised ^ontrolled trials wit a, hig ris of bias | **C** A body of eviden^e in^luding studies rated as + dire^tly appli^able to t e target population and de onstrating overall ^onsisten^y of results, or<br>E trapolated eviden^e fro studies rated as ++ |
| ++ Hig -quality sy ste ati^ reviews of ^ase-^ontrol or ^o ort studies or hig -quality ^ase-^ontrol or ^o ort studies wit a, very low ris of ^onfounding, bias or ^ an^e and a, hig probability t at t e relations ip is ^ausal | **D** Eviden^e level or , or<br>E trapolated eviden^e fro studies rated as + |
| + Well-^ondu^ted ^ase-^ontrol or ^o ort studies wit a, low ris of ^onfounding, bias or ^ an^e and a, oderate probability t at t e relations ip is ^ausal | **oo pr . po n** |
| - Case-^ontrol or ^o ort studies wit a, hig ris of ^onfounding, bias or ^ an^e and a, signifi^ant ris t at t e relations ip is not ^ausal<br>on-anal ti^al studies, e g ^ase reports, ^ase series<br>E pert opinion | ✓ ^e o ended best pra,ti^e based on t e ^lini^al e perien^e of t e guideline develop ent group |

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The guidelines review process will commence in unless evidence requires earlier review

DISCLAIMER

The Royal College of Obstetricians and Gynaecologists produces guidelines as an educational aid to good clinical practice The College presents recognised methods and techniques of clinical practice, based on published evidence, for consideration by obstetricians and gynaecologists and other relevant health professionals The ultimate judgement regarding a particular clinical procedure or treatment plan must be made by the doctor or other attendant in the light of clinical data presented by the patient and the diagnostic and treatment options available within the appropriate health services

This means that RCOG Guidelines are unlike protocols or guidelines issued by employers, as they are not intended to be prescriptive directions defining a single course of management Departure from the local prescriptive protocols or guidelines should be fully documented in the patient's case notes at the time the relevant decision is taken