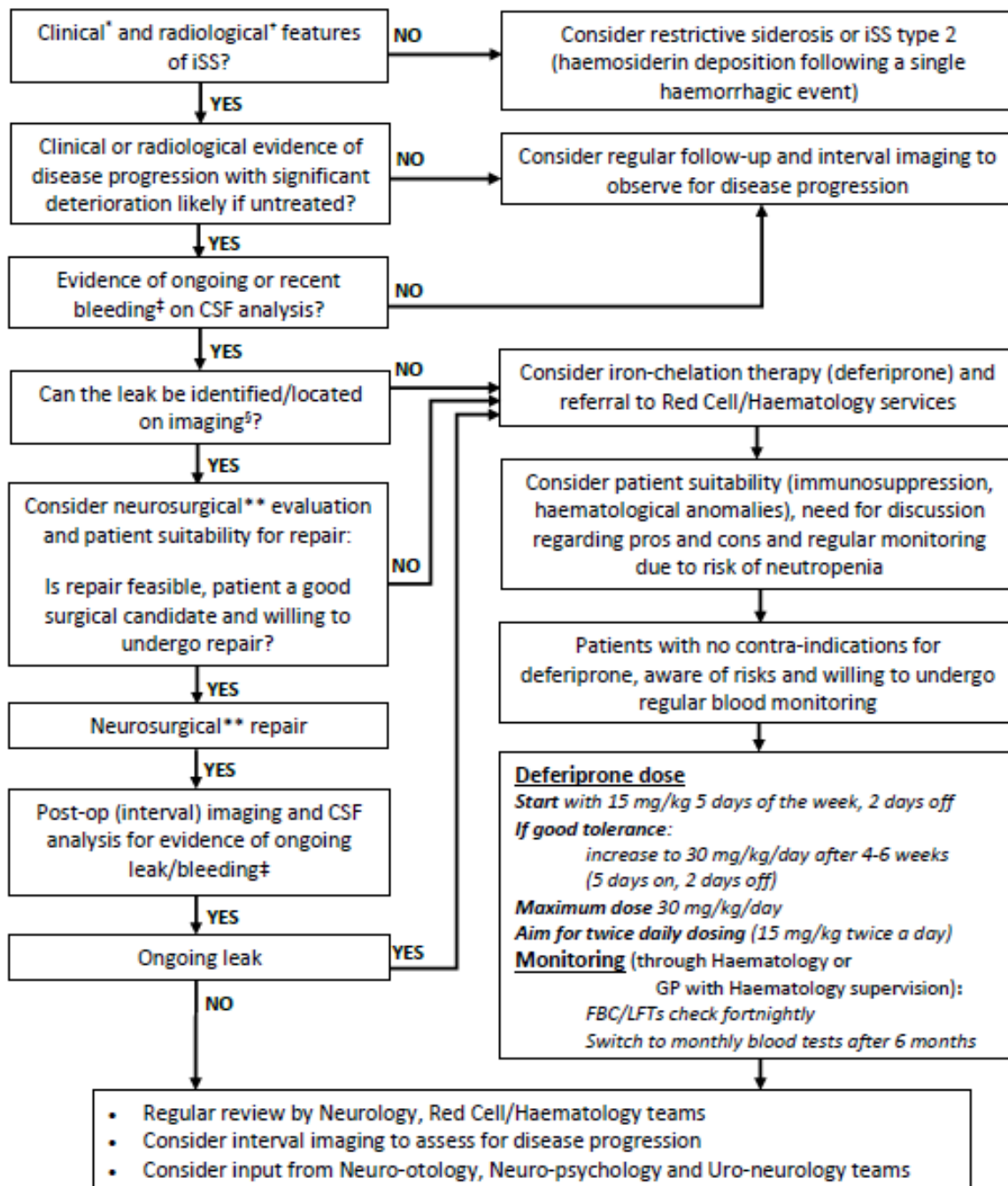


SUPPLEMENTAL MATERIAL

Flowchart of Queen Square iSS clinical care pathway



Legend:

*Common clinical features: hearing loss, ataxia/imbalance, myelopathy, sphincter disturbance, cognitive decline

‡Defined as: presence of haemosiderin in symmetrical distribution in at least 2 regions: (1) cerebellum (superior vermis, folia peduncles), (2) brainstem (midbrain, pons, medulla), (3) spinal cord or cranio-cervical junction

§Elevated CSF ferritin, presence of xanthochromia (bilirubin, oxyhaemoglobin), markedly elevated red-cell count (in high hundreds or thousands, unlikely due to a traumatic tap)

**Consider radiologically guided blood/glue patch as an alternative for patients who are otherwise considered poor surgical candidates (reduced functional reserve).

Supplemental figure 1. Flowchart of the proposed clinical care pathway.

Classical infratentorial superficial siderosis of the central nervous system: pathophysiology, clinical features and management

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Author, year	Cases (n)	Age in years, sex (M/F), (n, if case series)	Treatment offered (Y/N)			Follow up (months)	Clinical outcome (Y/N, n)			Radiological & CSF outcomes (Y/N, n)			
			Surgical repair (n)	Deferiprone			Improved	Stable	Worse	Resolution of bleeding into CSF, CSF collection or lesion excision	HS cranial appearance		
				Duration (months)	Side effects						Improved	Stable	Worse
Kumar ¹ , 2021	3	65 (mean), 2 females	3	N		N/R (2) 3 (1)	N/R (3)			N/R (2) 1	N/R (3)		
Meshkat ² , 2021	2	62 & 30, 2 males		18	N	18	2			N/R		2	
Nose ³ , 2021 & Egawa ⁴ , 2013	15	51 (mean), 6 males 55 (mean), 5 males	7 8	19 (mean)	N	19 (mean) 21 (mean)	7(o)		6(o)	Y^(o) Y^(o)	7(o)		7(o)
Schievink ⁵ , 2021	1	56 (M)	1	N		46	P			Y	Y		
Takai ⁶ , 2021	7	59 (median), 4 males	7	N		18 (median)		6	1	Y	N/R (7)		
Cornips ⁷ , 2020	1	33 (M)	1	N		18	P			P		Y	
Katoh ⁸ , 2020	1	74 (F)	1	N		12	P			Y		Y†	
London ⁹ , 2020	1	62 (M)	1	N		5	P			Y (Intra-op)	N/R		
Mohammed ¹⁰ , 2020	1	73 (M)	1	N		61	Y			Stable		Y	
Nathoo ¹¹ , 2020	1	74 (M)	1	5	N/R	8			Y‡	N/R	N/R		
Sammarraiee ¹² , 2020	10*	52 (mean), 7 males	2	28 (mean) 25 (mean)*	IDA (4) Neutropaenic sepsis (3)* Fatigue (1)* Joint pain (1)	28 (mean)		6	4	N/R		8*	
Sato ¹³ , 2020	1	65 (M)	1	N		12	Y			Y	N/R		
Teranishi ¹⁴ , 2020	1	16 (F)	1	N		12	Y			Y		Y	
Wiacek ¹⁵ , 2020	1	63 (M)	1	N		36		Y		N^	N/R		
Cossu ¹⁶ , 2019	4	64 (mean), 4 males		44 (mean)	N	44 (mean)	2	2		N/R	4		
Levy ¹⁷ , 2019	1	54 (M)		120	N/R	120		P		N/R	Y		
Machino ¹⁸ , 2019	1	71 (M)	1	N		"Post-op"		Y		Y	N/R		
Mbadugha ¹⁹ , 2019	1	50 (M)	1	N		12		Y		IMP^	N/R		
Vellutini ²⁰ , 2019	1	35 (M)	1	N		12	P			Y^	Y		
Arishima ²¹ , 2018	2	50 (M) 59 (M)	1 1	N N		17 9		Y		Y Y	N/R N/R		
Brembilla ²² , 2018	1	48 (M)	1	24	N	24	Y			Y/Y^	Y		
Camlar ²³ , 2018	1	58 (F)	1	N		12		Y		Y	Y		
Derle ²⁴ , 2018	1	63 (M)		9	N	9		Y		N/R	N/R		
Hosokawa ²⁵ , 2018	3	57 (mean), 3 males	3	N		24		3		Y/P^ (3)	N/R (3)		

						(4 in 1 case)							
Johans²⁶, 2018	1	10 (F)	1	N		12		Y		Y		Y	
Kessler²⁷, 2018	38**	64 (mean), 20 males	8	24 (mean)	Fatigue (8) Mild zinc deficiency (5), Neutropaenia (2), Joint pain (2), Mouth ulcers (2)	24 (Mean)	4	15	11	N/R	8		8
Espinoza Rodriguez²⁸, 2017	1	35 (M)	1	N		12		Y^^		P^^	N/R		
Kuo²⁹, 2017	1	59 (F)		6	N	6	Y			N/R	Y		
Madkouri³⁰, 2017	1	58 (M)	1^^^	N		N/R	N/R			N/R	N/R		
Sinha³¹, 2017	3	3 males (age N/R)	1 1 1	N/R‡	N/R	N/R 24 N/R	P P		Y‡	Y Y Y	N/R		
Takai³², 2017	1	58 (M)	1	N		12	Y			Y	N/R		
Bihari³³, 2016	1	56 (F)	1	N		24	Y			Y	N/R		
Decramer³⁴, 2016	1	60 (M)	1	N		12			Y	N/R	N/R		
Ryu³⁵, 2016	1	55 (M)	1	N		6			Y	N/R		Y†	
Takai³⁶, 2016	1	19 (M)	1	N		12	P			Y	N/R		
Kondziella³⁷, 2015	3	59 (mean) 2 males	1 2	N/R N		N/R N/R		Y	Y	Y^ (3)	N/R (3)		
Tosaka³⁸, 2015	1	69 (M)	1	N		12			Y	No recurrence	N/R		
Katoh³⁹, 2014	1	36 (M)	1	N		>1		Y		N/R	N/R		
Pikis⁴⁰, 2014	1	33 (M)	1	N		24		Y		N/R	N/R		
Saft⁴¹, 2014	1	48 (M)	1	N		>1	Y			N/R	N/R		
Schirinz⁴², 2014	1	67 (M)		3	N	6	Y‡			P		Y‡	
Yokosuka⁴³, 2014	1	53 (M)	1	N		12		Y		Y^	N/R		
Cummins⁴⁴, 2013	1	65 (M)		12	Transient nausea at onset of treatment	12	P			N/R		Y	
Huprikar⁴⁵, 2013	1	65 (M)		4‡	Neutropaenic sepsis‡	6	Y‡			N/R		Y	
Tari-Capone⁴⁶, 2013	1	20 (M)	1	N		"Post-op"		Y		N/R	N/R		
Boncoraglio⁴⁷, 2012	1	69 (M)	1	N		6		Y		Y/Y^	N/R		
Levy⁴⁸, 2012	10††	55 (mean), 5 males		3	Transient abnormal LFT (3)	3	4	4	2	N/R	4	3	2

Levy ^{49 50} , 2012 & 2011	1	65 (M)		38	Fatigue Joint and muscle aches Transient IDA	38	Y			N/R	Y		
Cheng ⁵¹ , 2011	1	53 (M)	1	N		6	P			IMP			N/R
Driver-Dunckley ⁵² , 2010	1	54 (M)	1	N		12	P			N/R			N/R
Gonella ⁵³ , 2010	1	67 (M)	1	N		9		Y		N/R			N/R
Ikeda ⁵⁴ , 2010	2	71 (F) 67 (F)	1 1	N N		12 8		Y Y		P^ Y^			N/R N/R
Kumar ⁵⁵ , 2010	1	54 (M)	1	N		4		Y		P	Y†		
Payer ⁵⁶ , 2010	1	47 (F)	1	N		24	Y‡‡			Y		Y	
Aoyama ⁵⁷ , 2009	1	36 (M)	1	N		3	Y			Y^			N/R
Hoxworth ⁵⁸ , 2009	1	53 (F)	1	N		3		N/R		Y			N/R
Kumar ⁵⁹ , 2009	1	64 (M)	1	N		6	Y			Y			N/R
Shih ⁶⁰ , 2009	1	70 (M)	1	N		15		Y		N/R			N/R
Holle ⁶¹ , 2008	1	59 (M)	1	N		6			Y	Y^			N/R
Levy ⁶² , 2007	1	48 (M)		24 (T)	N/R	24			Y	N/R			N/R
Spengos ⁶³ , 2007	1	63 (M)	1	N		12		Y		N/R			N/R
Konya ⁶⁴ , 2006	1	49 (F)	1	N		12	Y			Y			N/R
Kumar ^{65 66} , 2006 & 2005, Cohen-Gadol ⁶⁷ , 2004	7***	60 (mean) 6 males	7	N		16-24 (120***)	1	3	1***	Y(4)/Y^(2)			N/R (7)
Miliaras ⁶⁸ , 2006	2	45 (M) 44 (M)	1	N 24 (T)	N/R	24 24		No IMP		No IMP			
Aquilina ⁶⁹ , 2005	1	34 (M)	1	N		36		Y		N/R		Y	N/R
Kole ⁷⁰ , 2004	1	51 (M)	1	N		8		Y		Y^			N/R
Messori ⁷¹ , 2004	1	65 (M)	1	N		2		N/R†††		N/R			N/R
Kale ⁷² , 2003	1	35 (M)		36 (T)	N/R	36		Y		N/R			N/R
Leussink ⁷³ , 2003	2‡‡‡	63 (F) 52 (M)	1	N 24 (T)	N/R	7 24		Y	Y‡‡‡	N/R IMP^‡‡‡			N/R N/R
Das ⁷⁴ , 2001	1	50 (M)	1	N		8			Y^^	N^^			N/R
Haroun ⁷⁵ , 2000	1	47 (M)	1	N		6	Y			Y (Intra-op angiography)			N/R

Supplemental table 1. List of reported cases with classical infratentorial superficial siderosis describing iron-chelation therapy or surgical intervention (including Gamma-knife surgery for tumour resections) with curative intent/to repair source of bleeding, or both; clinical and radiological outcomes included. Side effects (or none) of iron-chelation therapy were included where reported. Literature search was limited to English and years 2000 - 2021; publications not included if describing: (1) debulking/palliation surgery, (2) theco- or ventro-peritoneal shunting; (3) dural blood patching (3) medical therapy other than iron-chelation.

Legend:

(o) Overall clinical and radiological outcomes for each group (number of participants at follow-up stated, Nose et al³)

^ Based on CSF analysis (at 6 month-follow up (Mbadugha et al¹⁹, Brembilla et al²², Vellutini et al²⁰); resolution of xanthochromia at 4-month follow-up but subsequent signs of rebleeding at 14 month follow-up, Wiacek et al¹⁵)

† Spinal MRI (at 3-month follow-up, Katoh et al⁸; at 2-month follow-up, Ryu et al³⁵; at 4-month follow-up, Kumar et al⁵⁵)

‡ Deferiprone stopped in view of significant clinical deterioration (Nathoo et al¹¹); deferiprone commenced due to progression of symptoms after surgical repair (Sinha et al³¹); clinical improvement and stable MRI haemosiderin appearances after 3-month therapy however clinical deterioration after stopping deferiprone (Schirinzi et al⁴²); clinical improvement during deferiprone which was stopped due to neutropaenic sepsis (Huprikar et al⁴⁵)

* Deferiprone discontinued due to treatment-related complications in 4 patients, interval imaging (available in 8 cases) performed 21 (mean) months after starting the medical treatment (Sammaraiee et al¹²)

** Post-treatment clinical data available in 30 of 38 participants; clinical outcomes assessed on 5 parameters and 3 categories: (1) hearing (better/same/worse in 0%/42%/58%); (2) coordination (better/same/worse in 6%/26%/68%); (3) walking (better/same/worse in 3%/29%/68%), (4) fine motor function (better/same/worse in 0%/29%/71%) and (5) bowel/bladder function (better/same/worse in 3%/39%/58%); there was no indication of better outcomes for 8 participants who had surgery than in non-surgical arm; post-treatment imaging available for 16 participants (Kessler et al²⁷)

^^ Tumour resection incomplete (Espinoza Rodriguez et al²⁸); dissemination leading to clinical decline (Das et al⁷⁴)

^^^ Clipping of dural arteriovenous fistula

>1 Follow-up stated as “several months” (Katoh et al³⁹); and “several weeks” (Saft et al⁴¹)

†† Interval (Mean=34 months) imaging reported in 9 cases (Levy et al⁴⁸)

‡‡ Clinical (ISS-related) improvement at 2-year follow-up, subsequent significant deterioration likely from dual pathology (amyotrophic lateral sclerosis) (Payer et al⁵⁶)

*** Follow-up time not reported in 3 cases; clinical outcomes not reported in 2 cases; resolution of collection/bleeding confirmed on CSF in 2 cases and not reported in another 2 cases; 1 patient

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showed progression/deterioration at 10 -y follow-up but it was unknown if bleeding was corrected at surgery (Kumar et al⁶⁵);

‡‡‡ Post-operative complication reported: bilateral subdural haematomas, resolving after 2 months (Messori et al⁷¹)

††† Partial clinical improvement recorded at 3-month follow-up and deterioration in clinical signs reported at 24-month follow-up; CSF exam at 4 months demonstrated resolution of RCC and protein, siderophages persisted (Leussink et al⁷³);

CSF = cerebrospinal fluid; DFP = deferiprone; F = female; HS = haemosiderin; IDA = iron deficiency anaemia; IMP = improved; Intra-op = intraoperative; LFT = liver function test; M = male; MRI = magnetic resonance imaging; N = no; N/R = not reported; P = partial improvement; Post-op = post-operative; T= trientine; Y = yes;

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