14/361

UK National Screening Committee

Screening for Fatty Acid Oxidisation Disease

19 March 2015

Aim

1. This document provides background on the item addressing newborn screening for Fatty acid oxidisation disorders.

Current policy

2. The current UKNSC policy recommends that newborn screening for the following fatty acid oxidisation disorders should not be included in the current newborn bloodspot screening programme:

- Carnitine uptake defect
- Long chain hydroxyacyl CoA dehydrogenase deficiency
- Trifunctional protein deficiency
- Very long chain acyl CoA dehydrogenase deficiency

3. This is based on a 2003 HTA study addressing a number of inherited metabolic disorders:

4. The recent evaluation of expanded newborn screening resulted in a recommendation not to introduce screening for long chain hydroxyacyl CoA dehydrogenase deficiency (LCHADD) or trifunctional protein deficiency (TFP). Therefore, the attached review document only considers the evidence on screening for very long chain acyl CoA dehydrogenase deficiency (VLCADD) and carnitine uptake defect (renamed as carnitine transporter deficiency (CTD).

Current Review

Bazian were asked to assess the literature published since the previous review, taking into literature from January 2001 to July 2013. The resulting document is attached.

5. The findings of the review identified these key areas:

- The clinical course of both VLCADD and CTD is variable, and there is no reliable way to predict phenotype/prognosis.
- There is uncertainty over the accuracy of the screening test as most screening studies have not performed extensive follow-up and therefore false-negatives could have been missed. For VLCADD, this is additionally complicated by the fact that it can be difficult to distinguish heterozygous carriers from affected cases in which mutation in only one allele can be found.
- Screening for both conditions can identify heterozygotes, and the natural history of heterozygotes is not well understood and
- Although there are accepted treatments for both conditions, there is uncertainty over whether all cases identified through screening will require treatment.

Consultation

6. A three month consultation was hosted on the UK NSC website between November 21st and February 23rd and additionally promoted through the PHE Screening Twitter platform. The following organisations were contacted directly: ALD Life, British Inherited Metabolic Disease Group, Children Living with Inherited Metabolic Diseases, Clinical Genetics Society, Genetic Alliance UK, Institute of Child Health, Rare Disease UK, Royal College of Paediatrics and Child Health, Save Babies Through Screening Foundation UK, UK Newborn Screening Laboratories Network, NHS

England Specialised Commissioning and the Department of Health rare diseases team.

7. Responses were received from Genetic Alliance UK, a joint response from the Patient Advocate for Newborn Screening (PANS) and Save Babies through Screening Foundation UK, Jim Bonham (on behalf of MetBioNet) and three families with experience of VLCADD.

8. More of the comments received focussed on the recommendation not to screen for VLCADD than for CTD. The Issues raised in the consultation responses for both VLCADD and CTD were:

- Broadly, it was acknowledged that there was evidence and knowledge gaps in the current understanding of both conditions.
- How the gaps in the evidence for both VLCADD and CTD could be closed before the UK NSC next reviewed screening for the conditions.
- Several responses stated that a lack of understanding of the natural history shouldn't undermine the benefit that they believed would be brought about by screening.
- That the most recent ENBS conditions being implemented were as a result of UK pilots to find out more information prior to screening and advocating this to be done for VLCADD.
- Other countries, including several EU member states all using the same appraisal criteria that screen for VLCADD suggesting that there was overall benefit of screening for this condition.
- The UK NSC having a preference for peer reviewed studies and not taking into account qualitative evidence from the patient voice in its decision making.
- Personal experiences from 3 families with children and a parent who have VLCADD who shared the impact that VLCADD has had on them. The dietary plans and emergency regimens from their respective hospitals were also provided along with their response.
- A query with regards to whether relevant studies were missed by the review (one of which, Engvall et al., 2010, describing mild forms of VLCADD that presented after long bouts of exercise) was published before the literature search cut-off). This study

suggested universal newborn screening would have benefited the three mild cases by advising them not to undertake long bouts of vigorous exercise. The study did not discuss how an early prognosis advocating mild prevention rather than more extensive treatment options, like absence of fasting, would be arrived at following newborn screening.

- Xxxxxxxx xxxxxxxxx comments on both reviews included:

    - A study by Spiekerkoetter et al.,2009 that showed no deaths from 20 screen detected cases but two death from 10 clinically detected cases confirmed the benefit of screening for VLCADD.
    - An acknowledgment that VLCADD did not satisfy criterion 11 (*agreed evidence based policies covering which individuals should be offered treatment and the appropriate treatment to be offered*) but the point was put that i) this was also true for other metabolic disorders screened to a lesser or greater extent ii) whether the lack of clarity regarding the treatment needed for some should prevent access to asymptomatic screening detection and treatment where clearly indicated for others.
    - Several European screening programmes for VLCADD suggesting there was a simple, safe and validated screen test for VLCADD, with the variability in cut-offs for screening of conditions not invalidating the test. Additionally, the possibility of using the same cut-off as Netherlands, whose blood spot screen is at the same time point as the UK.
    - That, while genotype-phenotype correlations were poor, clinical indicators, metabolite studies, enzyme assay and flux studies would enable sufficient personalised patient plans to be developed.
    - The cost and acceptability of screening for VLCADD being in line and possibly improved by the existing of the current blood spot programme.
    - A belief that the recommendation not to screen for VLCADD may be too conservative based on the evidence of benefit.

The full consultation responses can be found in Annexe A.

9. Following on from the consultation comments a meeting will be held between Genetic Alliance UK and the Director of the UK NSC, Anne Mackie, to discuss the issues relating qualitative review evidence.

FMCH March 2015 Meeting

10. The FMCH approved the following recommendation at its March meeting:

    "The UK NSC does not support changing its current recommendation of not screening for VLCADD and CTD fatty acid oxidisation disorders.

Action

11. The UK NSC is asked to approve the above recommendation

UK National Screening Committee
Screening for Fatty-acid Oxidisation Disorders - an evidence review
VLCADD

Consultation comments

Name:	**Jim Bonham**	**Email address:**	Xxxxxxxxxxxxxxxx
Organisation (if appropriate):	MetBioNet		
Role:	**Laboratory Director**		

Do you consent to your name being published on the UK NSC website alongside your response?

Yes ☒ **No** ☐

Section and / or page number	Text or issue to which comments relate	Comment *Please use a new row for each comment and add extra rows as required.*
P11	Table 5	The evidence shown is convincing in that 2/10 clinically detected cases died whereas 0/20 screen detected cases died

P14	Second bullet point	The favourable outcome in screen diagnosed cases (second bullet point) supports a benefit from screening
P20	Criterion 2 conclusion	It is therefore difficult to conclude that the natural history is insufficiently clear to recognise the benefit conferred by screening. It is true that not everything is known but there is sufficient evidence and understanding to expect benefit if not complete clarity.
P28	Criterion 5 conclusion	It is clear from the 12+ countries that offer screening that a simple and safe test exists validated by extensive practice over many years.
P36	Criterion 6 conclusion	Cut-offs as in all screening vary country to country but this does not invalidate the test. The Netherlands has a similar time of testing and cut-offs here may offer a guide.
P38	Criterion 7 conclusion	I am not aware of studies exploring the acceptability of the test for the other metabolic disorders that form part of the programme but the test and the issues for VLCADD are similar and no reason to believe that the test would be less acceptable in this context
P39	Criterion 8 conclusion	While it is accepted that genotype-phenotype correlation may be poor, this is true for other disorders. In practice it would be possible by considering clinical indicators, metabolite studies, enzyme assay and flux studies to design personalised patient plans and this is common practice when treating IMDs.
P43	Criterion 10 conclusion	In practice patients would be assessed on an individual basis depending on the clinical circumstances and the laboratory findings, the broad principles of treatment are clear for this disorder and the evidence suggest that they are effective.
P44	Criterion11 conclusion	While this is true, it is also true for other metabolic disorders screened to a lesser or greater extent. The treatment needed and benefits will be clear for many patients but less clear for

		others, the question is whether the lack of clarity for some should prevent access to asymptomatic recognition and treatment where this is clearly indicated for others
P44	Criterion 13 conclusion	This criteria cannot be achieved for rare heterogeneous conditions at this stage but it does not invalidate screening
P44	Criterion 14 conclusion	There is no real reason to doubt this based on the experience of screening for other rare IMDs
P46	Table 19 estimated marginal costs	The marginal start-up costs for the other four recently added IMDs is known, £0.59, and adding VLCADD would be very unlikely to exceed this figure in a UK context
P46	Criteria 18, 19 and 20.	While not assessed these are inherently no more demanding than for other IMDs and therefore achievable.
P48	Conclusions	The conclusions are probably a bit too conservative, it is clear from the evidence that screening for VLCADD can be beneficial, while it is accepted that it is a heterogeneous condition, like many IMDs and indeed other screened disorders, screening for this disorder can benefit a significant number of patients with acceptable specificity and sensitivity. The choice of treatment is likely to be determined on an individual patient basis and will be less intrusive in equivocal cases. Practical screening cut-offs and diagnostic testing could be put in place for the UK population at our time of testing.

UK National Screening Committee
Screening for Fatty-acid Oxidisation Disorders - an evidence review

Consultation comments

Name:	**Pat Roberts**	**Email address:**	Xxxxxxxxxxxxxxxxxxx
Organisation (if appropriate):	Save Babies Through Screening Foundation UK (SBUK) and Patient Advocates for NBS Group (PANS)		
Role:	**Executive Director of SBUK and Chair of PANS**		

Do you consent to your name being published on the UK NSC website alongside your response?

✓ **Yes** ☐ **No** ☐

Section and / or page number	**Text or issue to which comments relate**	**Comment** *Please use a new row for each comment and add extra rows as required.*
General Comment	Full Report	We support robust evidence, however currently 9 EU Member States screen for VLCADD. All EU member states apply the same set of criteria when determining whether screening is appropriate, criteria is met and the benefits to the patient will do more good than harm. Therefore 9 Member states cannot all be screening newborns in a reckless manner. Some of the issues raised in the policy review are issues that are therefore appropriate to be considered and resolved for the UK population in pilot studies. The recommendation to retain the status quo and not screen for VLCADD or introduce a pilot study cannot therefore be supported from the perspective of

		patient/public voice.
General	Full Report	We have had the opportunity to see and consider the responses made by Prof Jim Bonham in response to this policy review document. We fully support the observations made by Prof Bonham. There is no purpose in repeating these excellent points and have therefore commented on additional points or evidence that is key to the patient perspective.
Page 11, Table 5 Page 48	Page 11, Table 5 Clinical presentation of 30 patients with VLCADD reported in Spiekerkoetter et al. (2009) Conclusions	There is evidence that screening for VLCADD can be beneficial. In table 5 the evidence shows that 2 out of 10 clinically detected cases died, whilst no cases out of 20 detected through NBS died As with many IMDs, screening for this disorder can benefit a significant number of patients. The choice of treatment is likely to be determined on an individual patient basis and will be less intrusive in equivocal cases.
Page 48	Conclusion Screening test	Practical screening cut-offs and diagnostic testing could be put in place for the UK population at our time of testing.
Page 15 Page 48	Conclusions	It is difficult to understand why the natural history is insufficiently clear to recognise the benefit conferred by screening.
Page 50	Conclusions: Implications for research.	The policy review by the UK NSC team has identified a lack of evidence in a number of areas. The review has identified where studies in 4 main areas may assist in driving out the necessary evidence to support screening for VLCADD in the UK. However no suggestion is made by the UK NSC of what work might be done and by who to obtain this evidence. Without suggestions, on next steps the policy will be boxed forward for review in another few years and the same conclusion will be reached. Something concrete needs to happen to review and provide the necessary evidence

General	Additional published evidence not identified and perhaps not considered.	Early diagnosis and treatment of the fatty acid oxidation disorder VLCADD in patients identified by newborn screening (Eskens FJM, Jones I, Lutyen K, University Hospital Antwerp. Journal of Inherited Metabolic Disease, September 2014. Fatty Acid Oxidation Defects revealed by Extreme Physical Activity, Case Report with Implications for Newborn Screening (Engvall, Barbara, Wibom, Nennesmo, Bieneck Haglind, von Dobeln) Journal of Inherited Metabolic Disease August 2010. False positive rates in NBS of MCAD and VLCAD deficiency scheduled on 5th day after birth. (Kagawa, Tsmura,Hara, Satoshi, Tajima, Sakura, Hata, Shigematsu, Kobayashi) Journal of Inherited metabolic disease September 2013. I noticed that there are other published findings within the Journal of IMD for VLCADD that have not been included in this policy review. However some of these cover distinct populations e.g. Saudi Arabia. Is it the policy of the UK NSC to disregard evidence where research is unlikely to impact a UK population, or have these just been missed?

UK National Screening Committee
Screening for Fatty-acid Oxidisation Disorders - an evidence review

Consultation comments

Name:	**Alastair Kent**	**Email address:**	Xxxxxxxxxxxxxxxxxxx
Organisation (if appropriate):	Genetic Alliance UK is the national charity working to improve the lives of patients and families affected by all types of genetic conditions. We are an alliance of over 180 patient organisations. Our aim is to ensure that high quality services, information and support are provided to all who need them. We actively support research and innovation across the field of genetic medicine. Rare Disease UK is a multi-stakeholder campaign run by Genetic Alliance UK, working towards the delivery and implementation of a national strategy for rare diseases in the UK. The UK Strategy for Rare Diseases was published in November 2013. Pertinent to this consultation, the Strategy includes a commitment from all four Governments of the UK to: "Continue to work with the UK National Screening Committee to ensure that the potential role of screening in achieving earlier diagnosis is appropriately considered in the assessment of all potential new national screening programmes and proposed extensions to existing programmes." Commitment 9, The UK Strategy for Rare Diseases, November 2013. This commitment recognises the value that the rare disease community places on early diagnosis, not only for the benefits it can bring to an affected individual but because of the impact it can have on improving the quality of life for their whole family.		
Role:	**Director**		

Do you consent to your name being published on the UK NSC website alongside your response?

Yes X **No ☐**

Section and / or page number	Text or issue to which comments relate	Comment *Please use a new row for each comment and add extra rows as required.*
General	Overreliance on published literature for an evidence review	The current methodology used by the UKNSC when making decisions about whether the benefits of introducing a newborn screening programme for a condition outweighs the risks places a premium on peer reviewed literature to the exclusion of all other forms of evidence. Relying solely on peer reviewed literature excludes the direct contribution of the patient voice to the process. While information from clinicians and patients may not be published, it represents the most recent and relevant information on a condition coming from those that either directly manages or is affected by the condition today. Not taking this type of information into account during a review of the evidence is out of step both with other institutions with responsibility for decisions regarding public health, such as NHS England, the National Institute for Health and Care Excellence and the European Medicines Agency, and with accepted practice in dealing with rare disease issues. All three of these agencies, and more, have accepted that evidence will always be scarce in the area of rare disease, and is likely to be of weaker statistical significance than that expected from more common conditions. They have resolved to fill this gap by accepting qualitative evidence from the patient community. We believe the UK NSC should take steps to do the same. As the national organisation representing those affected by inherited conditions, Genetic Alliance UK would welcome a

		meeting to discuss where we could assist in this process.
Screening for Very Long-Chain AcylCoenzyme A Dehydrogenase Deficiency		
Page 50	"Implications for research - Outcome studies, especially of asymptomatic infants detected by screening. - Studies to determine whether phenotype or outcome can be predicted - Studies to determine the optimal management of patients with VLCADD, and who needs treatment - Studies of the natural history of heterozygotes to determine whether they are at risk of disease, especially those heterozygotes with VLCAD activities similar to cases of VLCADD"	Genetic Alliance UK recognise that there are significant gaps in knowledge about VLCADD, the link between phenotype and prognosis, the benefits of early dietary intervention and the clinical relevance of being a heterozygous carrier of a VLCADD related mutation. The UK NSC's review highlights the absence of peer reviewed and published evidence on these areas, as well as follow-up studies on patients diagnosed through newborn screening While it is clear that a better understanding of the areas highlighted by the UK NSC's review would be valuable, what is not clear is how this information is likely to be generated within a reasonable time frame. The UKNSC only considers evidence that has been published in a peer reviewed journal, and favours those studies that specifically look at patients in the UK and in the context of the UK healthcare system. Given these limitations, it is unlikely that the types of evidence that the UKNSC rely on using to inform their decisions will be produced without proactive work by the UKNSC and associated stakeholders. We note that of the 47 publications referenced in this current review nearly three quarters are dated from prior to 2010, with 23% more than ten years old.

		The last four conditions that were added to the newborn screening programme (homocystinuria, maple syrup urine disease, glutaric aciduria type 1 and isovaleric acidaemia) were included following a pilot where these conditions were screened for routinely at birth in a small number of centres. Without the evidence gathered by this pilot, it would not have been possible for the UK NSC to satisfy their evidence requirements and positively recommend newborn screening for these conditions. We would encourage the UKNSC to consider establishing a similar pilot for VLCADD and related conditions in order to address this. As VLCADD is already part of newborn screening programmes in the USA and eleven European countries, it is likely that the pilots would be successful and provide the UKNSC with sufficient evidence to support the introduction newborn screening for VLCADD in the UK, particularly as a dietary treatment for this condition is already available.

UK National Screening Committee
Screening for Fatty-acid Oxidisation Disorders - an evidence review

Consultation comments

Name:	**Alastair Kent**	**Email address:**	Xxxxxxxxxxxxxxxxxxx
Organisation (if appropriate):	Genetic Alliance UK is the national charity working to improve the lives of patients and families affected by all types of genetic conditions. We are an alliance of over 180 patient organisations. Our aim is to ensure that high quality services, information and support are provided to all who need them. We actively support research and innovation across the field of genetic medicine. Rare Disease UK is a multi-stakeholder campaign run by Genetic Alliance UK, working towards the delivery and implementation of a national strategy for rare diseases in the UK. The UK Strategy for Rare Diseases was published in November 2013. Pertinent to this consultation, the Strategy includes a commitment from all four Governments of the UK to: "Continue to work with the UK National Screening Committee to ensure that the potential role of screening in achieving earlier diagnosis is appropriately considered in the assessment of all potential new national screening programmes and proposed extensions to existing programmes." Commitment 9, The UK Strategy for Rare Diseases, November 2013. This commitment recognises the value that the rare disease community places on early diagnosis, not only for the benefits it can bring to an affected individual but because of the impact it can have on improving the quality of life for their whole family.		
Role:	**Director**		

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Yes X **No ☐**

Section and / or page number	Text or issue to which comments relate	Comment *Please use a new row for each comment and add extra rows as required.*
General	Overreliance on published literature for an evidence review	The current methodology used by the UKNSC when making decisions about whether the benefits of introducing a newborn screening programme for a condition outweighs the risks places a premium on peer reviewed literature to the exclusion of all other forms of evidence. Relying solely on peer reviewed literature excludes the direct contribution of the patient voice to the process. While information from clinicians and patients may not be published, it represents the most recent and relevant information on a condition coming from those that either directly manages or is affected by the condition today. Not taking this type of information into account during a review of the evidence is out of step both with other institutions with responsibility for decisions regarding public health, such as NHS England, the National Institute for Health and Care Excellence and the European Medicines Agency, and with accepted practice in dealing with rare disease issues. All three of these agencies, and more, have accepted that evidence will always be scarce in the area of rare disease, and is likely to be of weaker statistical significance than that expected from more common conditions. They have resolved to fill this gap by accepting qualitative evidence from the patient community. We believe the UK NSC should take steps to do the same. As the national organisation representing those affected by inherited conditions, Genetic Alliance UK would welcome a

		meeting to discuss where we could assist in this process.
Screening for Carnitine Transporter Deficiency		
Page 46	"Implications for research Further research is required into: - The UK prevalence/incidence of CTD - Follow-up studies of asymptomatic infants and mothers detected by screening - Whether phenotype/prognosis can be predicted - The natural history of heterozygotes - The timing of newborn specimen collection to avoid maternal carnitine levels influencing test results"	Genetic Alliance UK recognise that there are significant gaps in knowledge about CTD, including the number of children affected, the link between phenotype and prognosis and the clinical relevance of being a heterozygous carrier of a CTD related mutation. The UK NSC's review highlights the absence of peer reviewed and published evidence on these areas, as well as on the practical elements of introducing a screening programme (the timing of sample collection and follow-up studies after diagnosis through screening). While it is clear that a better understanding of the areas highlighted by the UK NSC's review would be valuable, what is not clear is how this information is likely to be generated within a reasonable time frame. The UKNSC only considers evidence that has been published in a peer reviewed journal, and favours those studies that specifically look at patients in the UK and in the context of the UK healthcare system. Given these limitations, it is unlikely that the types of evidence that the UKNSC rely on using to inform their decisions will be produced without proactive work by the UKNSC and associated stakeholders. We note that of the 38 publications referenced in this current review a third dated from prior to 2010, with 20% more than ten years old.

		The last four conditions that were added to the newborn screening programme (homocystinuria, maple syrup urine disease, glutaric aciduria type 1 and isovaleric acidaemia) were included following a pilot where these conditions were screened for routinely at birth in a small number of centres. Without the evidence gathered by this pilot, it would not have been possible for the UK NSC to satisfy their evidence requirements and positively recommend newborn screening for these conditions. We would encourage the UKNSC to consider establishing a similar pilot for CTD and related conditions in order to address this. As CTD is already part of newborn screening programmes in the USA and seven European countries, it is likely that the pilots would be successful and provide the UKNSC with sufficient evidence to support the introduction newborn screening for CTD in the UK, particularly as a treatment for this condition is already available.

UK National Screening Committee
Screening for Fatty-acid Oxidisation Disorders - an evidence review
CTD deficiency

Consultation comments

Name:	**Jim Bonham**	**Email address:**	Xxxxxxxxxxxxxxxxxxx
Organisation (if appropriate):	MetBioNet		
Role:	**Laboratory Director**		

Do you consent to your name being published on the UK NSC website alongside your response?

Yes ☒ **No** ☐

Section and / or page number	Text or issue to which comments relate	Comment *Please use a new row for each comment and add extra rows as required.*
P4	Treatment	A lack of RCTs for rare disease treatment does not imply a lack of efficacy but rather the rarity of the conditions
P6	Criterion 1	It is difficult to conclude, if the condition can be fatal that it is not serious
P24	Criterion 2 conclusion	It is accepted that this is a heterogeneous disorder without a clear genotype phenotype correlation in which asymptomatic adult patients are described equally the early detection afforded by screening appears to confer benefit for some individuals and no fatalities have been described in early detected patients treated before the onset of symptoms.

P37	Criterion 6 conclusion	Cut-offs as in all screening vary country to country but this does not invalidate the test.
P40	Criterion 10 conclusion	Again the lack of RCT evidence in this context does not imply a lack of effectiveness and this is an important principle
P42	Criterion 13 conclusion	A lack of RCT evidence is not a relevant consideration for rare disorders
P43	Table 24 estimated marginal costs	The marginal start-up costs for the other four recently added IMDs is known, £0.59, and adding VLCADD would be very unlikely to exceed this figure in a UK context
P44	Criteria 18, 19 and 20.	While not assessed these are inherently no more demanding than for other IMDs and therefore achievable.
P48	Conclusions	While it is compelling that no fatalities have been reported in screen identified individuals it is accepted that when compared with other fat oxidation defects such as MCADD and LCHADD that the case for screening is less convincing, largely due to the numerous reports of asymptomatic adults and the heterogeneity of the condition. The decision to screen then becomes a value judgement about whether offering life changing benefits to some individuals in a way that only screening can achieve outweighs medicalising others who may never require treatment

XXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXXX

DEPARTMENT OF NUTRITION AND DIETETICS

DIET PLAN

NAME: xxxxxxx		Home
HOSPITAL NUMBER:	xxxxxxxx	DATE: xxxxxxx

MAXIMUM FASTING TOLERANCE 6 HOURS Needs 4.6-7.7g of CHO per hour Max 6g of long chain fat from food per day 0.8g of long chain fat from Key Omega 1250mls fluid a day	
7:30: BREAKFAST (23g or more of CHO) 150ml Monogen + 1 sachet of Key Omega	
10:30am: Mid-morning snack (20g or more of CHO)	
1pm: Lunch (20g or more of CHO)	
3:30pm Mid afternoon snack (20g or more of CHO)	
5:30pm:TEA (15g or more of CHO per meal)	
MID EVENING and NIGHTIME	
6:30pm:150ml of Monogen (27g or more of CHO) Overnight feed via gastrostomy (65g or more of CHO) 62mls/hour 12.5% Polycal solution over 8 ½ hours 10:00pm -6:30am	
DIETITIAN: xxxxx xxxxx (Student dietitian) Xxxxx xxxxxxx	Mob: xxxxxxxxxxx

xxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxx

xxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxxx

Name: xxxxx xxxxxxx Date: xxxxxxxx

Emergency Regimen 20% Carbohydrate
2 -10 years
For use when unwell

Recipe
Small volume recipe 40g or 8 level scoops of Polycal using scoop provided in the tin. Make up to 200ml with water. **Large volume recipe** 200g or 40 level scoops of Polycal using scoop provided in the tin. Make up to 1000ml with water. **S.O.S 20** Empty sachet of S.O.S 20 into bottle Make up to 200ml with water **Oral rehydration solution recipe** 1 sachet of Dioralyte or Electrolade or Rapolyte Add 30g **or** 6 level scoops of Polycal **or** 1 sachet of S.O.S 20 Make up to 200ml with water **Recipe if using commercial drinks (NOT Sugar Free)** Look at the nutritional label per 100g: Examples of suitable commercial drinks: Fruit Juice Fruit Shoot and Fruit Shoot 100% (all flavours) Carton Ribena – (original blackcurrant, strawberry, apple) Fizzy Drinks eg: Coca Cola, Fanta, Sprite, 7UP, Pepsi Regular **If commercial drink has 9 to 12g carbohydrate per 100ml:** Add 10g or 2 level scoops of Polycal **If commercial drink has 13 to 16g carbohydrate per 100ml:** Add 5g or 1 level scoop of **Polycal**
1. Use level scoops to measure out powder. 2. Try to make up feeds one at a time if possible as this reduces risk of infection. 3. Store made up feeds in a refrigerator and throw away 24 hours after making them up 4. Always thoroughly wash hands and make feeds up in a clean environment.

Amount to offer:
Suggested Drink Volumes:

Age 2 years: aim 1200ml in 24 hours
Offer: 100ml every 2 hours or 150ml every 3 hours day and night

Age 3-4 years: aim 1300ml in 24 hours
Offer: 110ml every 2 hours or 170ml every 3 hours day and night

Age 5-6 years: aim 1500ml to 1600ml in 24 hours
Offer: 130ml every 2 hours or 200ml every 3 hours day and night

Age 7-8 years: aim 1700ml in 24 hours
offer: 135ml every 2 hours or 210ml every 3 hours day and night

Age 9 years: aim 1800ml in 24 hours
offer: 150ml every 2 hours or 220ml every 3 hours day and night

Paediatric Metabolic Dietetic Team: xxxxxxxxxx/ xxxxxxxxxxxxxxx
Contact Number xxxxxxxxxxxxx (answerphone)/ xxxxxxxxxxx

xxxxxxxxx

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xxxxxxx

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xxxxxxxx

My personal experience by xxxxx xxxxxx pg 1

I was born on the xxxx xxxxxxxx xxxxx a normal birth although a very fast delivery as mum and dad said "I popped out like a cork". My parents can't quite remember the exact age but between four to six months old I was taken very poorly and rushed to accident and emergency in a critical condition, after tests they found that my liver and brain was swollen and was continuing to do so. They gave me phenobarbitone to try and stop the swelling and other tests which I am unaware of. After surviving this critical time I was diagnosed with Reyes syndrome which we now know was a misdiagnosis. The next ten years I spent in and out of hospital, my parents say it felt like every couple of weeks I was on a glucose drip. This had some good results and they then added hypoglycaemia to the diagnosis. Mid to late teens gave a little respite from hospital but with no ryme or reason to why but I just managed to recover at home with glucose tablets and rest, although I was a lot less active in these years compared to a young child running around all day. Illness such as

Personal experience continued by xxxxx xxxxxx pg2

Colds and viruses still have the same effect as it did in my younger days to what it does now, but unfortunately with a much more impact.

After several years of this happening my mum decided it was time to see the GP, who diagnosed growing pains but I knew deep down this was more than that but who was gonna listen to a teenager?

By the time I left school and started full time work as a health care assistant at my local hospital I never really experiences pain but I felt tired all the time. As I was 18 years old now I just put it down to my body not used to lots of hours and hard work, but I did eventually go to the GP again and get tested but they only tested for anaemia which was negative. I decided I was just unfit and got on with life like this and throughout my 20 ' s I felt what I could only explain as normal. I worked as a student nurse in my 20's tile was 24 years old when I qualified and took on a full time job at my local hospital as a staff nurse where I stayed til my youngest son xxxxx was 8 months old. I had become a district nurse when I was 26 which I loved but found the bending up and down on my knees so hard and painful, but I didn't really think anything of it as all my colleagues complained too. I left this job role for a more relaxed environment role as a practice nurse at my local gp surgery, where I stayed til I went

on maternity leave at 29 years old to have my second son **xxxxx** and this is where it all started.

Personal experience continued by xxxxx xxxxxx pg 3

xxxxxx was 4 weeks old and I went shopping on this particular day to my local town to get some bits ready for my 30th birthday which was coming up, after 3 shops I felt sick, shaky and in a lot of pain all over and I knew I needed to get home as I thought it was the start of flu.

I called the nursery to collect my oldest son xxxxxx and we all got into bed and this is where we stayed til my husband xxxxx came home. Xxxxx was made redundant this day and although it was tough it was also good timing as the months ahead were very hard.

After spending 2 weeks at home in bed and seeing a gp at home in that time who diagnosed a urine infection as my urine was gravy in colour and no antibiotics made any difference. I began to feel better after a week but my legs appeared very weak so my husband took me to see my GP who run some tests. I had an abnormal liver blood test and was then refered to a rhumatologist at my local hospital who diagnosed a virus, but several episodes later they decided to refer me to xxxxxxxx hospital as this was more serious than a virus.

Xxxxxxxxx run a few tests including skin and muscle biopsy which in 7 months I finally got a diagnosis of very long chain

acyl Coa dehydrogenase deficiency (VLCAD) a very rare genetic metabolic myopathy where my body is unable to convert certain fats to energy.

Personal experience continued by xxxxxx xxxxxx pg 4

It appeared the trauma of my son's birth had triggered this and that I've had this from birth, which now explains all my symptoms as a newborn through to adult hood which Reyes syndrome we now know is a mistake diagnosis for vlcad.

2012 I was made retired due to ill health from my practice nurse job role and after several relapses I now use a mobility scooter/wheel chair and have adaptations at home and my husband xxxxxx is my full time carer as each relapse has caused severe muscle damage and weakness.

My experience of VLCAD by xxxxxx xxxxx pg 1

The last 3 years have been very challange for me and my family with big life style changes and we are still everyday facing new challange. At present I am currently under the care of **xxxxx** hospital for a clinical trial.

I feel early detection of vlcad is vital as it can prevent muscle damage and weakness leading to life needing mobility aids and a better quality of life in general, therefore I am fighting alongside xxxx xxxxxx mum to xxxxxx 5 with vlcad and xxxxx (angel) 2 years as well as xxxx xxxx mum to xxxxx 4 years with vlcad needing peg feeds at night to have vlcad added to the newborn screening system to give each newborn baby a chance in life without struggling with their quality of life.

At present we are currently raising awareness and organising a family fun day for the spring/summer.

Thank you for reading

Xxxx xxxxx xxxxx wife/mum to xxxxx, xxxxx & xxxxx.

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xxxxxxx

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Personal experience of VLCAD from xxxxxx xxxxxx on behalf of 2 sons xxxxx & xxxxx

xxxxxxx was born at 11.21pm he never cried and was wide awake. He didn't seem interested in feeding but as the midwives requested I kept trying to breastfeed. He slept majority of the night, still not interested in feeding. The following morning as the doctors did the ward round I told him how he hadn't been feeding much and I couldn't give them an estimate of ounces due to breastfeeding. He was a little floppy on observation. To me he just seemed like a delicate content newborn. They took him up to Neonatal where he was treated for hypothermia. He had a low temperature, blood sugars were low and blood work had started to be done. 'Was this my fault? Did I not wrap him up properly during the night?' was all that was going round in my head at this point. I continued with the breastfeeding until I thought it would be best to go onto a bottle so we could see what he was taking. After a few days we were discharged and went home.

A little over a week at home we got a call from the hospital to go straight in. They advised me to stop feeding xxxx and come immediately. The doctors were waiting for us at the door as we arrived and explained that the bloodwork they had done had come to show some abnormality. xxxxx had a metabolic condition called Very Long Chain Acyl-CoA Dehydrogenase Deficiency (VLCAD) an inherited metabolic disorder of long chain fat oxidation. Because of this xxxxx is unable to use fat as an energy source during times of fasting or intercurrent illness or prolonged exercise.

From here it was arranged that we would see a genetics team and metabolic specialist at xxxxxxx Childrens Hospital. Everything happened so fast, he had a skin biopsy, more blood tests and myself and my husband, xxxxxxx, were diagnosed as carriers of this gene. We were given a diet plan and medication information for when xxxxx was older and the treatments we would need to follow.

xxxx has spent majority of his life in and out of hospital being treated with relapses of the condition. So much so in July 2013 he had a portacath fitted due to his veins being very weak and hard to access during emergencies. A portacath is an implanted venous access device which is fitted underneath the skin. He began with muscle weakness at around 8 months. He didn't crawl he just dragged himself along the floor. As he began walking shortly after his first birthday we noticed signs of him having pains in his legs and was admitted regularly for this. His Creatine Kinase was always raised.

In November 2013, xxxxx had his most severe relapse. His blood sugars dropped so quickly and he became very lethargic and unable to rouse, he then had a hypoglycaemic seizure. Following this he needed a lumbar puncture and head CT scan. As he came round he had slightly slurred speech and was uneasy on his feet for a few days.

I am unable to count the amount of times xxxxx has been admitted but now he is 5 years old things are starting to get easier. He understands that when he feels ill he needs to tell us straight away.

In August 2012, I had my second child xxxxxx. As we knew the risks we were on hand with special low fat formula and a plan in place with Neonatal following his delivery. He also had some low blood

Sugar issues but fed well. We stayed in Neonatal until the genetics test came back which revealed that he too was positive for VLCAD. As we knew how to deal with it this time round we got on with life following xxxxxx plan and watching for signs of illness. Very rarely xxxxx needed hospital intervention. He was the complete opposite to his brother. He was walking at 10 months and never had any muscle problems. In September 2014, xxxxx started with a sickness bug, he was admitted to hospital just to be on the safe side due to the lack of fluids. After one night he was discharged and back to his usual self for the rest of the day. The following morning we went to wake xxxxx as he was usually up before us, he was very pale and his breathing was shallow. I checked his blood sugars and instantly rang an ambulance. As they arrived xxxx had stopped breathing, he was rushed to hospital where he had to be resuscitated several times. Due to the hypoglycaemia, xxxxxx had severe swelling on his brain. The hospital did everything they could but within a few hours of trying they explained that the damage to his brain was devastating and beyond repair. We then had to endure another 24 hours of agony waiting for a brain stem test to reveal what we already knew.

This is my personal experience of VLCAD. Until starting a social media page I knew of nobody else with this condition, even doctors had to ask me twice what it was. I now have people share their experiences with me, if we need any advice and support or just a general chat. Unfortunately though, the vast majority of these people are overseas and hardly any in the UK. We need more awareness out there as it is so severe especially in newborns.

Xxxxx xxxxx

Xxxxxx xxxxxx

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xxxxxx

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xxx xxx

Personal experience of VLCAD from xxxx xxxxxx on behalf of son *xxx xxxxx* age 4.

xxxxx was born on xxxxxxxxx *xxxxx* at 6.30pm at *xx xxxxx* hospital *xxxxxxx* following a completely normal and happy pregnancy. There were no concerns either from us or the midwives.

I was induced at 42 weeks and following a slow progressing delivery the decision was made for xxxxx to be born by caesarean section. A perfectly healthy baby, just a little on the cold side and had to have a heater on him for some time following delivery.

xxxxx breastfed without concern and seemed to have a normal appetite, the nurses on the ward were happy with his progress and had he not been born by c-section we'd have probably been able to be discharged home.

On the second day in the afternoon *xxxxx* seemed a bit unsettled and was crying and seemed to be in discomfort but we put this down to wind and did not have any further concerns.

Being a first time parent I had decided to follow the advice in a book called the contented little baby to try and establish an early feeding and sleeping routine as obviously these little gifts do not come with their own set of instructions! One of the suggested feed times was for 1.30am and I was awake with him and watching him waiting for this time to arrive, finally when it did I first went to change him. I changed his nappy and was doing his sleep suit back up when I noticed he was not moving, I put my face by his and could not detect breath, I gently rubbed his chest and called his name but nothing. I picked him up and ran down the ward where a midwife took him from me and in to another room where the crash team were called.

xxxxxxx had suffered a cardio respiratory arrest and after resuscitation was taken to NICU where he suffered further seizures and his glucose had dropped to 1.3. We were told at this point that there was a chance he would not make it or that he would likely have some brain damage and we were all left wondering what had happened to this happy, healthy little boy.

We were so fortunate in that at xx xxxxxx hospital there was xxxxxx xxxxxx xxxxxx, one of the founders of cooling therapy and it was decided that xxxxx fitted the criteria for this and wheels were put in to motion. He was put in an induced coma and had a

cooling jacket on bringing his body temperature down and slowing his brain function. On initial observations his brain pattern was all over the place and showing

Signs of extreme abnormalities but then soon after the therapy started we had the wonderful news that his brain was 'normalising'. After 72 hours he was slowly warmed back up and finally we were soon able to hold our little boy.

Following the therapy xxxxx started to receive my expressed breast milk but was also still receiving IV fluids but seemed to be recovering well, when it became time to remove his ventilator we all crossed our fingers and for a while all seemed to be going well but he then suffered a relapse and was put back on it.

At 9 days old xxxxx was suitably recovered to have an MRI which showed mild watershed type hypoxic ischaemic injury, plus a small bleed was discovered. We do not know exactly how this damage will manifest, Bailey III examination where he scored in a lower range than for his age.

At 17 days old we were finally allowed to take our little boy home. All of the tests had returned a negative result, he was tested also for sleep apnoea and while there were some respiratory issues there was no cause for major concerns. We were told that there were still some results outstanding for some of the rarer conditions.

At home xxxxx continued to feed well but in between he was unusually sleepy and often difficult to rouse, in comparison to other babies he was around he barely opened his eyes.

After about a week at home we received a call from Dr xxxxx xxxxxxat xxxxx Childrens Hospital confirming a positive result for VLCAD and I was to stop breastfeeding him immediately and we were admitted on to the ward where we were to meet his metabolic team and so began his special low fat diet.

We were reviewed at Clinical Genetics and both myself and my husband found to be carriers. We were told at the time that they had found 2 changes to xxxx gene one of which was mild and the other unknown, though the following confirmation letter advised 2 tiny changes both having been seen before in other patients.

Diet and Feeding

xxxxxx was initially fed on a 3 hourly basis and at 10 months old had a gastrostomy to enable overnight feeding. He had a fasting test at 1 year old and the pre-meal bloods that were taken had improved and so he was able to go to 4 hours, he was tested again the following year and tested for up to 7 hours and while his blood glucose remained ok it was discovered once the blood results were back that fats had started metabolising and it was not safe to go that long and so he remains on 6 hour fasting. We have been advised that he is likely to require his peg until 8 years of age when he may be able to take a cornstarch supplement. He currently is allowed up to 7 grams of fat per day with one being taken up by key omega. He requires 8 grams of carbohydrates per hour for his current weight.

VLCAD presentations

xxxxx has so far returned a normal ECG aside from a small duct which has not closed over from birth and being reviewed in one years time with a view to possibly operate.

His CK levels are checked at each review and so far found to be normal.

His energy levels are generally high and he does not show signs of pain or discomfort following exercise.

xxxxx really only displays VLCAD signs in illness where he can deteriorate alarmingly quickly even in mild illness. He spent his first 2 and a half years in and out of hospital with various bugs and illnesses but fortunately aside from visits related to his mini button (gastrostomy) we have been incident free for 18 months or so.

Awareness and information

Whilst his metabolic Doctor, dieticians and nurse have been absolutely fantastic it has only been in this last year that I have found other families in the UK living with VLCAD and information is very hard to come by. Considering the severity of the condition and massive benefit of early management giving the best chance of not just survival but also of avoiding irreversible symptoms it is surprising that there is not more awareness about it, especially in this day when we hear more and more about mitochondrial disease in general. I hope that can change soon as currently we find ourselves being the expert over the doctor that is treating him.

The power of knowing you’re not on your own and meeting others in a similar situation has been the best therapy so far.

[redacted] Children's Hospital NHS Foundation Trust

Department Nutrition & Dietetics NHS

MINIMAL FAT DIET

FOODS ALLOWED

Meat & meat substitutes
Lean Red meat (5g fat/100g or less)*
eg beef, pork, lamb
Soya mince, Quorn*, tofu*

Poultry
Chicken* (white breast meat, no skin)
Turkey *(white breast meat, no skin)

Fish
White Fish (no skin), eg haddock, cod, plaice

Shellfish eg crab, crabsticks prawns, shrimps, lobster

Milk and Milk Products
Skimmed milk (fresh or dried)

Very low fat yoghurts or fromage frais

Low fat soft cheese. Quark
Low fat Cottage cheese

Bread & crackers
French bread – unlimited
White bread*, wholemeal bread*

Rye crispbread. Rice Cakes, Matzos

FOODS NOT ALLOWED

Meat & meat substitutes
Fatty meats such as belly pork, breast of lamb
Sausages, salami, fat on meat
Meat pies, burgers
Pate, potted meat

Poultry
Chicken/Turkey (dark meat or skin)
Chicken/Turkey in batter/breadcrumbs/sauces/pastry
Duck, goose

Fish
Oily fish eg salmon, mackerel
Herrings, sardines, kippers
Fried Fish
Fish in batter/breadcrumbs/sauces

Milk and Milk Products
Whole milk/ semi skimmed milk (fresh or dried)
Dried milk with added vegetable fat eg Five Pints, Pint Size
Evaporated milk, condensed milk
Cream
Full fat/whole milk/low fat yoghurts
Thick n Creamy yoghurts
Ice Creams
Full and half fat hard or soft cheese
Ordinary soft or cottage cheese

Bread & crackers
Other bread *
Naan bread, chapatti made with fat
Croissant
Oatcakes, cheese crackers, crackers

FOODS ALLOWED

Pasta, Rice & Grains
Pasta –all types except wholemeal pasta*

Noodles
White rice (basmati best). Brown rice*
Couscous, bulghur wheat

Flours and cereals
White flour, wholemeal flour*
Corn flour, custard powder
Semolina, sago, tapioca

Breakfast Cereals
Breakfast Cereals- most are suitable
Wholewheat types, eg Weetabix, Bran Flakes are higher in fat than non wholewheat types eg Rice Krispies, Corn Flakes

Cakes & Biscuits & Pastry
Only those made from low fat ingredients
95% fat free cakes and biscuits*

Desserts/ Puddings
Skimmed milk puddings eg rice, custard

Jelly, meringue, water ices, sorbet

Sugars & Preserves
Sugar, honey, jam, marmalade, golden syrup, treacle

Confectionary
Boiled sweets, mints, jelly sweets, fruit gums
Fruit pastilles, marshmallow, plain ice lollies

Soups,
Clear soup, consommé, very low fat soups
Some low calorie and 'healthy eating type' soups are very low fat

Sauces & Gravies
Marmite, Oxo, Bovril, very low fat gravy mixes & stock cubes
Brown sauce, tomato ketchup
Very low fat dressings/mayonnaise
Soy sauce, tomato puree
Sauces made with skimmed milk and corn flour
Minimal fat sauces (jars/tins/packets)

FOODS NOT ALLOWED

Pasta, Rice & Grains
Pasta in dishes eg macaroni cheese, carbonara
Egg noodles
Yorkshire Puddings, Dumplings

Flours and cereals
Soya flour
Bran. Foods made with flour which contain fat eg pastry, cake

Breakfast Cereals
Cereal with nuts eg muesli
All Bran, Ready Brek

Cakes & Biscuits & Pastry
Cakes, biscuits, buns, pastry for sweet and savoury foods

Desserts/Puddings
Puddings made with whole or semi skimmed milk or with fats eg butter
Sponges- all types. Fritters

Sugar & Preserves
Chocolate spread, lemon curd
Marzipan

Confectionary
Chocolate, chocolate – covered sweets, fudge, toffee

Soups
Most soups, cream soups

Sauces & Gravies
Gravy made with fat, most gravy mixes & stock cubes

Salad Cream, mayonnaise

Other sauces
Sauce mixes (jars/tins/packets)

FOODS ALLOWED	FOODS NOT ALLOWED
Seasonings Salt, pepper, herbs, spices Vinegar, pickles & chutneys	**Seasonings**
Beverages Tea, coffee, Horlicks, Ovaltine, drinking chocolate Cocoa- all made using skimmed milk Fruit Juice, fruit squash/cordial Fizzy drinks Milk shake syrups	**Beverages** Complan Drinks made with whole or semi skimmed milk
Fruit and Vegetables All fruit, vegetables, salad except opposite (Fresh, frozen, tinned, dried) Very Low fat crisps*	**Fruit and Vegetables** Avocado Pear, Olives Coleslaw, Potato Salad Fried Vegetables Potatoes - roast, chips Crisps, low fat crisps
Pulses Peas eg chick peas, split peas, lentils Beans eg red, white, borlotti, black - eyed	**Pulses**
Eggs Egg white	**Eggs** Egg yolk
Nuts & Seeds	**Nuts & Seeds** Nuts, peanut butter, seeds eg sesame, sunflower
Fats and Oils MCT Oil, as permitted	**Fats and Oils** Butter, margarine, low fat spread Vegetable oils, lard, suet, dripping

Please Note:

- Foods marked * may need to be restricted because of their fat content. You will be advised by the dietitian as needed
- MCT Oil is not suitable for use by everyone – please ask your dietitian for advice
- Some foods in the 'avoid list' have very low fat alternatives to the regular high fat foods which can be included in the diet eg sauces, desserts, ice creams, cheeses

[redacted] CHILDREN'S NHS FOUNDATION TRUST NHS
DEPARTMENT OF NUTRITION AND DIETETICS

METABOLIC EMERGENCY REGIMEN – AMOUNT & CONCENTRATION OF GLUCOSE POLYMER REQUIRED AT DIFFERENT AGES

Age	Carbohydrate solution (% or g per 100ml)	Glucose Polymer+Water Dilution	Daily Volume
0 - 3 months	10	2 scoops up to 100ml or 1 sachet SOS 10 made up to 200ml	45-60ml every 2 hours or 70-120ml every 3 hours
4 - 6 months	10	2 scoops up to 100ml or 1 sachet SOS 10 made up to 200ml	85-100ml every 2 hours or 130-150ml every 3 hours
7 – 9 months	10	2 scoops up to 100ml or 1 sachet SOS 10 made up to 200ml	90-100ml every 2 hours or 130-150ml every 3 hours
10– 12 months	10	2 scoops up to 100ml or 1 sachet SOS 10 made up to 200ml	100ml every 2 hours or 150ml every 3 hours
1 – 2 years	15	3 scoops up to 100ml or 1 sachet SOS 15 made up to 200ml	100ml every 2 hours or 150ml every 3 hours Aim 1200ml in 24 hours
2 years	20	4 scoops up to 100ml or 1 sachet SOS 20 made up to 200ml	100ml every 2 hours or 150ml every 3 hours Aim 1200ml in 24 hours
3 and 4 years	20	8 scoops up to 200ml or 1 sachet SOS 20 made up to 200ml	110ml every 2 hours or 170ml every 3 hours Aim 1300-1400ml in 24 hours
5 and 6 years	20	8 scoops up to 200ml or 1 sachet SOS 20 made up to 200ml	130ml every 2 hours or 200ml every 3 hours Aim 1500-1600ml in 24 hours
7and 8 years	20	8 scoops up to 200ml or 1 sachet SOS 20 made up to 200ml	135ml every 2 hours or 210ml every 3 hours Aim 1700ml in 24 hours
9 years	20	8 scoops up to 200ml or 1 sachet SOS 20 made up to 200ml	150ml every 2 hours or 220ml every 3 hours Aim 1800ml in 24 hours
10 years	25	10 scoops up to 200ml or 1 sachet SOS 25 made up to 200ml	150ml every 2 hours or 220ml every 3 hours Aim 1800ml in 24 hours
11,12 and 13 years	25	10 scoops up to 200ml or 1 sachet SOS 25 made up to 200ml	170ml every 2 hours or 250ml every 3 hours Aim 2000ml in 24 hours
14 and 15 years	25	10 scoops up to 200ml or 1 sachet SOS 25 made up to 200ml	180ml every 2 hours or 270ml every 3 hours Aim 2200ml in 24 hours
16,17 and 18 years	25	10 scoops up to 200ml or 1 sachet SOS 25 made up to 200ml	200ml every 2 hours or 300ml every 3 hours Aim 2400ml in 24 hours

ALL DRINKS MUST BE GIVEN EVERY 2- 3 HOURS DAY AND NIGHT
The drinks can be sipped slowly over each 2 or 3 hours if not tolerated

[redacted] Children's Hospital NHS Foundation Trust
Department Nutrition & Dietetics NHS

TIPS FOR MINIMAL FAT COOKING & EATING

Cooking

- Remember to cook without adding fat – grill, braise, boil, stew, bake, casserole, steam or microwave instead of frying
- Base most of your meals on dishes made with pulses (dhal), white fish, shell fish, chicken/turkey
- Choose very lean cuts of meat and limit portion sizes
- Take the skin off chicken and turkey

Nutritional labelling

- Manufactured foods have nutritional labelling with total fat content expressed as grams of fat per 100g and sometimes also grams of fat per portion. If you have to restrict your fat to a set amount each day these can be used to calculate the amount of food that provides one gram of fat. This makes it easier to monitor and calculate your daily fat intake. Please ask your dietitian for more information if needed
- Commercial baby foods:
  'Wet' baby foods with 0.5g fat/100g or less and dried baby foods with 2g fat/100g or less are allowed freely.
  One serving per day allowed of 'wet' baby foods 0.5-1g fat/100g or dried baby foods 2-5g fat/100g
- For older children & adults:
  Manufactured foods which have a fat content of less than 0.5g per 100g can generally be allowed in the diet freely, unless large amounts are being consumed. Foods that contain 3g fat or less per 100g are considered to contain only a little fat. However these should still be taken in moderate amounts only
- There are many and potentially misleading words used to describe the fat content of food eg reduced fat, low fat, virtually fat free, 90% fat free. Ideally you should choose only foods labelled very low fat or virtually fat free

[redacted]

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