MODIFIED ATKINS KETOGENIC DIET - OUTPATIENT MODEL OF CARE FOR PATIENTS WITH EPILEPSY - SCH

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- The ketogenic diet (KD) is a high fat, adequate protein and low carbohydrate diet used under medical and dietitian supervision as a treatment for children with intractable epilepsy and certain metabolic conditions such as Glucose Transporter Type 1 (Glut1) deficiency.
- There are two types of KDs commonly used at Sydney Children's Hospital (SCH), the classical KD and the modified Atkins ketogenic diet (MAKD).
- The MAKD is often used for children over 2 years of age at SCH as it is a more palatable version of the KD, and is usually commenced in the outpatient setting, unless there is an indication to commence in the hospital.
- This practice guideline is intended for commencing and monitoring of MAKD in Outpatient setting only.
- The ketogenic diet is a medically supervised diet because it may impact growth and have other significant side effects including gastrointestinal intolerance, aspiration pneumonia, nephrocalcinosis and micronutrient deficiencies that need close medical, nursing and dietetic supervision.
- Due to the complexity and side effects of the KD, follow up is recommended 3 monthly in the outpatient setting for monitoring and evaluation, including growth, biochemistry, tolerance and compliance to the diet.

Approved by:	SCHN Policy Procedure and Guideline Committee	
Date Effective:	1st May 2021	**Review Period**: 3 years
Team Leader:	Staff Specialist	**Area/Dept**: Neurology

CHANGE SUMMARY

- The introduction has been updated to include the results of a Cochrane review of the ketogenic diet and other dietary therapies for epilepsy
- Updated procedure for requesting metabolic testing for ketogenic diet patients
- We have updated the flowchart documenting the MAKD patient pathway based on our current practice
- We have updated the medical, nursing and dietetic task lists for the pre-commencement, commencement and follow-up visits based on our current practice – these processes have been streamlined since the inception of the outpatient pathway for the MAKD
- Gastrointestinal complications (vomiting, diarrhoea, abdominal pain, anorexia) and aspiration pneumonia are well-recognised but infrequent adverse clinical events in patients receiving the Modified Atkins ketogenic diet[2]. We have modified the medical tasks at each clinical review to assess for gastrointestinal intolerance and risk of aspiration.
- We have modified the hypoglycaemia protocol to be in line with the most recent changes to the guideline: Hypoglycaemia on the ketogenic diet – SCH Practice Guideline
- The recipe for carbohydrate free potassium citrate (1.9 mmol potassium/ml oral liquid) has been included as an appendix.

READ ACKNOWLEDGEMENT

- This document is relevant for any clinicians at SCH who work in the area of the KD for children with intractable epilepsy/Glut1 deficiency syndrome. This includes:
  - medical staff
  - nursing staff
  - dietitians
  - pharmacists
  - neuropsychologists
  - epilepsy educators
  - theatre staff

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Related documents

- Dietitian referral form for ketogenic diet (including a list of contraindications)
- Hypoglycaemia on the ketogenic diet – SCH Practice Guideline.
- Protocol for minimisation of calculi risk associated with ketogenic diet
- Bone health in ketogenic diet patients

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Approved by:	SCHN Policy Procedure and Guideline Committee	
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1 Introduction

Epilepsy in which seizures persist, and seizure freedom is unable to be attained with anticonvulsant medication, is described as drug resistant, refractory or intractable epilepsy. Intractable epilepsy is also defined as the occurrence of one or more seizures per month over 18 months, despite treatment with at least two tolerated and appropriately dosed anti-epileptic drugs (AEDs) used either as monotherapies or in combination[1, 2]. Intractable epilepsy significantly increases the morbidity and mortality of those afflicted with epilepsy[3].

The ketogenic diet is a high fat, adequate protein and low carbohydrate diet used under medical and dietitian supervision as a treatment for children with intractable epilepsy or with inborn errors of metabolism including Glut1 transporter deficiency and some mitochondrial disorders.

The use of the ketogenic diet for epilepsy was first described in 1921[4]. The ketogenic diet needs to be medically supervised as there are many side effects and complications that need to be carefully monitored arising from administering a high fat and low carbohydrate diet to infants and children[5].

A Cochrane systematic review of the ketogenic diet and other dietary treatment published in 2012 using data from four randomised studies of the ketogenic diet found that in children, the ketogenic diet results in short to medium term benefits in seizure control, the effects of which are comparable to modern antiepileptic drugs[10].

There are currently four types of ketogenic diet. The two types used at Sydney Children's Hospital (SCH) are:

- Classical ketogenic diet
- Modified Atkins ketogenic diet (MAKD)

The classical ketogenic diet is the original form of the diet and is also the strictest. The classical ketogenic diet is expressed as a ratio of the amount of fat to protein and carbohydrate combined. This requires food to be weighed out on scales to the nearest gram.

The modified Atkins ketogenic diet (MAKD) was developed to be a less intensive and more palatable version of the ketogenic diet. Globally and internationally there has been a trend towards the use of the MAKD as efficacy has been shown to be comparable to the classical ketogenic diet[6] and has less risk of developing hypoglycaemia, which makes this form of diet more suitable to be administered as an outpatient [6, 7]. Unlike the classical ketogenic diet, the MAKD diet is not expressed in a ratio and food is not weighed or measured, and protein intake is not necessarily restricted. However, carbohydrate intake is usually restricted to 15g per day, and foods high in fat are encouraged and need to be included in every meal. For both the classical ketogenic diet and modified Atkins ketogenic diets, carbohydrate content in all oral and intravenous medications are counted in the total carbohydrate intake.

Aim

The aim of this guideline is to assist clinicians with the commencement and monitoring of the modified Atkins ketogenic diet for children with intractable epilepsy in an outpatient setting.

Scope

- Medical staff
- Nursing staff
- Dietitians
- Pharmacists
- Neuropsychologists
- Epilepsy educators
- Theatre staff

2 Patient selection

Children who are over 2 years and particularly school-aged children are usually commenced on a modified Atkins ketogenic diet.

The current practice at Sydney Children's Hospital is that infants (less than 2 years) and/or children receiving nutrition support via a feeding tube are commenced on the classical ketogenic diet as inpatients.

A screening process is conducted prior to starting any form of ketogenic diet, during which the patient is evaluated by medical staff to ensure there are no absolute or relative contraindications. An appropriate ketogenic diet is selected following discussions amongst the ketogenic team, the treating medical team, and the parents. Medical contraindications to commencing a ketogenic diet are listed below [8].

Absolute contraindications: Inborn Metabolic or inborn error/disorders of metabolism (IEMs)

- Carnitine deficiency (primary)
- Carnitine palmitoyl transferase (CPT) I or II deficiency
- Carnitine translocase deficiency
- Beta-oxidation defects
- Medium-chain acyl dehydrogenase deficiency (MCAD)
- Long-chain acyl dehydrogenase deficiency (LCAD)
- Short-chain acyl dehydrogenase deficiency (SCAD)
- Long chain 3-hydroxyacl-CoA deficiency
- Medium chain 3-hydroxyacl-CoA deficiency
- Pyruvate carboxylase deficiency
- Some mitochondrial disorders such as respiratory chain genetic disorders such as Complex IV (cytochrome C oxidase) and Complex V (ATP Synthase) deficiency
- Porphyria
- Prolonged QT interval or other cardiac diseases

Relative contraindications:

- Inability to maintain adequate nutrition due to gastrointestinal pathology or other causes
- Risk of aspiration pneumonia
- Surgical focus identified by neuroimaging and video EEG monitoring
- Parent or caregiver non-compliance
- Propofol concurrent use (risk of propofol infusion syndrome may be high)

For INFANTS, additional absolute and relative contraindications for commencing ketogenic diet also include the following, but are not discussed in details in this guideline[9]

Absolute

- Glycogen storage diseases (except type 2)
- Ketolysis defects
- Ketogenesis defects
- Liver, kidney or pancreatic insufficiency
- Hyperinsulinism

Relative

- Severe gastrointestinal reflux
- Familial hypercholesterolemia

A suitable patient for MAKD to be commenced in an outpatient setting

- Age 2 years and above
- Children with intractable epilepsy
- On an oral diet (caution for children with fussy eating behaviours)
- Family able to commit and attend allocated appointments
- Health care resources available to support families

Epilepsy Diet Blood Set on eMR:

Full blood count, electrolytes, urea, creatinine, calcium, magnesium, phosphate, liver function tests, albumin, selenium, zinc, 25-hydroxy-vitamin D, vitamin B12 (Total), Active B12, folate, iron studies, fasting glucose, triglycerides, cholesterol, total and free carnitine, acyl carnitine profile, plasma amino acids, Urine Calcium/Creatinine ratio, Urine Citrate/Creatinine ratio, betahydroxybutyrate

Urine metabolic screen is only done once as a screening test prior to commencing the diet (Request form must state that an inborn error of metabolism is suspected)

For results of total and free carnitine, acyl carnitine profile and plasma amino acids are sent to Children's Hospital Westmead (CHW) Biochemical Genetics, call 02 9845 3654 for results.

Request forms MUST document that the patient is commencing or on the ketogenic diet in order for CHW laboratory to perform investigations (plasma amino acids, carnitines)

PLEASE NOTE: total and free carnitine, acyl carnitine profile, plasma amino acids are very expensive tests and if ordered externally, the pathology service will charge the patient's family.

To find the Epilepsy Diet blood set and order them for a patient on eMR:

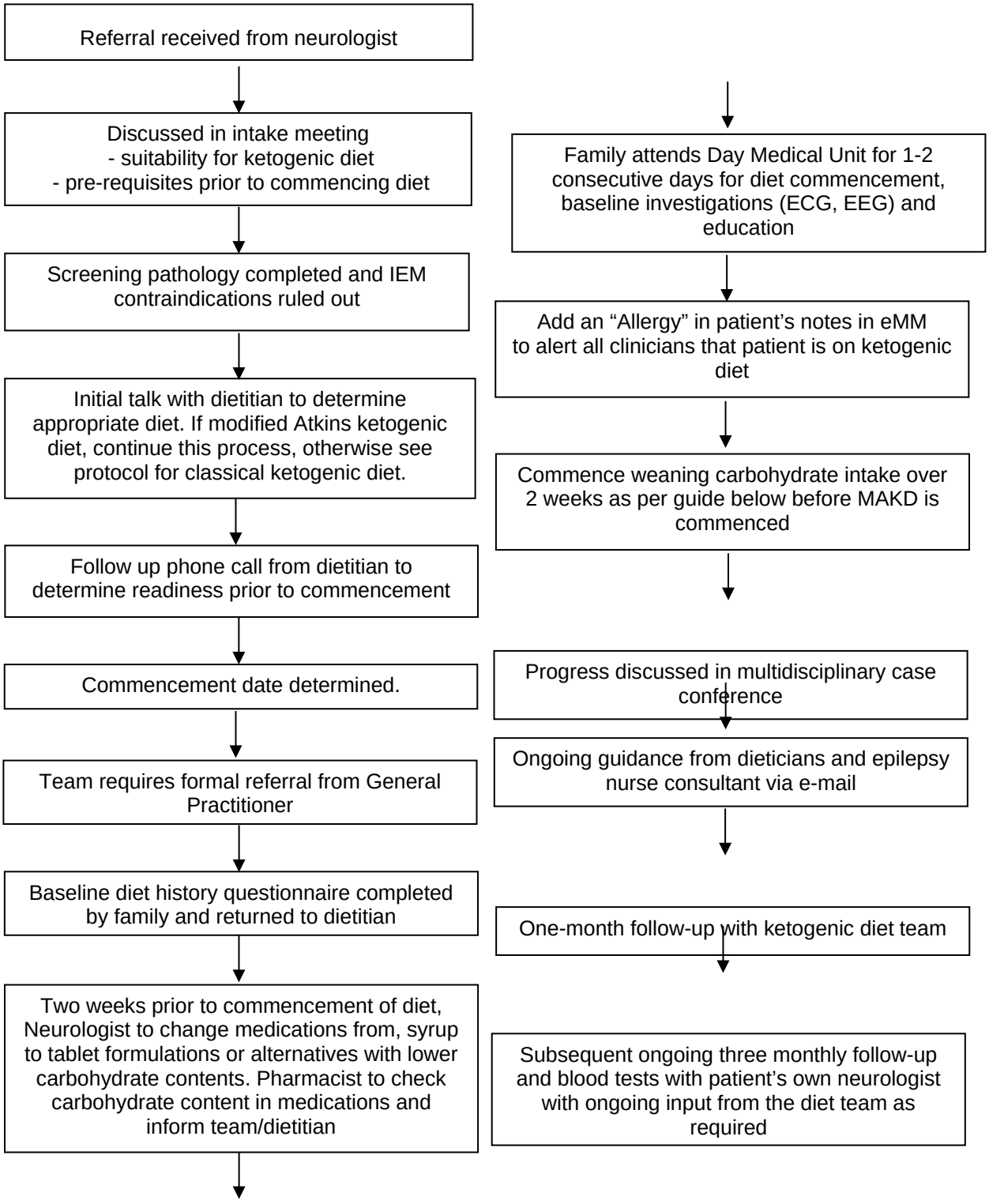
- Open up the patient medical record on Powerchart

- Click on “orders’ on side bar and press + add

- In the search dialogue box type in either “Epilepsy Order Set (SCH)” or “Neurology Epilepsy (SCH)”

- Entire Epilepsy Blood order set will automatically appear and can be selected.

Pathway for a typical patient initiated on the MAKD



Pre MAKD Commencement

Education provision

Location: Outpatient Clinic/Telephone consultation

Aim: To provide education and counselling for the Modified Atkins ketogenic diet to prepare for commencement of the diet

Tasks	Responsibilities
☐ Ensure screening pathology results are normal and contraindications are ruled out ☐ Assess risk for gastrointestinal intolerance and micro-aspiration ☐ Discuss practicalities of being on the MAKD and feasibility ☐ Discuss realistic expectations for seizure control and reductions in AEDs ☐ Medications to be changed to tablets or lower carbohydrate contents wherever possible	Medical staff
☐ Ensure baseline blood screening was arranged and attended prior to commencement of the MAKD and reviewed by Neurologist ☐ Report and seek medical advice on all results outside set parameters ☐ Ensure readiness to commence the diet and trouble shooting ☐ Book admission to medical day unit in liaison with secretary	Epilepsy CNC
☐ Conduct full nutrition assessment, including anthropometry parameters (weight, height, BMI and head circumference if appropriate) and estimate daily caloric requirements with a three-day food diary, likes and dislikes ☐ Assess suitability for MAKD ☐ Confirm carbohydrate content in medications ☐ Provide education on following and provide relevant written information: ☐ Background of ketogenic diet: efficacy, how it works ☐ Practicalities of ketogenic diet ☐ Short and long term side effects of ketogenic diet	Dietitian
☐ Confirm carbohydrate content in medications (give pharmacy at least 48-72 hours notice prior)	Pharmacist

Commencement of MAKD

Attendance over one-two days for diet commencement, education and baseline investigations

Location: Ambulatory Care Unit/Medical Day Unit

Aim: To provide further education and counselling to commence modified Atkins ketogenic diet at no more than 15g carbohydrate/day

Tasks	Responsibilities
☐ Check baseline pathology as per ketogenic diet set above (especially Total and Free Carnitine and Acyl-carnitine profile and amino acid levels for inborn errors of metabolism) ☐ Assess risk of gastrointestinal intolerance and micro-aspiration ☐ Document anthropometry parameters (weight, height, and head circumference if appropriate) ☐ Discuss possible side effects and management ☐ Educate parents on signs and symptoms of dehydration, acidosis, hypoglycaemia and nephrocalcinosis/kidney stones ☐ Review hypoglycaemia protocol and sick-day management protocol with family ☐ Ensure correct prescriptions have been written for community pharmacies (Pharmaceutical benefits scheme (PBS) Authority) as well as for the hospital pharmacy as indicated (Allow at least 72 hours notice to pharmacy) ☐ Ring GP/Paediatrician regarding commencement of ketogenic diet, discuss the need for sugar/carbohydrate free medications, sick-day management plans and monitoring of side effects ☐ Arrange follow-up in 1 month with ketogenic diet team in liaison with secretary ☐ Add onto the patient's allergy section on the eMM - "patient on ketogenic diet" (refer to Appendix 6).	Medical staff
☐ Attend baseline ECG and/or EEG ☐ Discuss monitoring of ketones and BGLs during transition to MAKD. Discuss mode for daily reporting of results ☐ Education on management of hypoglycaemia ☐ Ensure parents or carers have been educated on how to use the Freestyle Optium Neo BGL monitor and have received guidelines on safe use of the monitor, or assist in referral to HITH nursing staff if home follow up is required. ☐ Ensure parents or carers have been instructed on how to monitor urinary ketones with Keto-Diastix in the home setting and/or blood ketones with Freestyle optimum Neo monitor. Provide guidelines on the use of the monitor. ☐ Provide parents or carers with information on where to obtain consumables ☐ Provide troubleshooting and sick-day plan and letter for day care or school ☐ Educate parents or carers on signs and symptoms of dehydration and provide written resources ☐ Check that the patient's allergy section on the eMM - "patient on ketogenic diet" (refer to Appendix 6	Epilepsy CNC
☐ Check weight, height, BMI ☐ Provide further education on following and provide relevant written information: - Macronutrient intake - Dietary fats - Carbohydrate counting - Label reading ☐ Meal planning: calculate number of fat exchanges required per day and work out a plan for carbohydrate distribution ☐ Useful tools and resources (e.g. demonstration of how to use *Easy Diet Diary* or *Calorie King* app) ☐ Practical tips for preparing ketogenic meals	Dietitian

☐ Discuss short and long term side effects of ketogenic diet ☐ Recommend multivitamin and mineral supplements ☐ Organise samples of ketogenic diet specific formulas or supplements as appropriate	
☐ Review medications and confirm carbohydrate content in medications	Pharmacist

Introduction of low carbohydrate MAKD meals

Location: Home

Aim: Safely wean carbohydrate intake prior to reaching target of carbohydrate allowance (15g/day) over a period of 2 weeks

Process of weaning carbohydrate intake:

- MAKD is to be introduced over a period of 2 weeks (or longer if there are concerns with low BGLs).
- Introduce one MAKD meal (low carbohydrate meal) per day every 3-4 days (or longer if needed).
- The order of introduction of a MAKD meal needs to be discussed with parents. If children are attending school, suggest starting with breakfast or dinner.
- Carbohydrate is eventually reduced to 10-15g per day (including carbohydrate in medications) when MAKD is fully established.

Monitoring blood glucose levels (BGLs)

- Overnight or early morning BGLs are to be checked daily, usually starting when introducing the second or third MAKD meal, or if there are signs or symptoms of hypoglycaemia. BGLs also need to be checked for the first two days when fully established on the diet at 15g carbohydrate/day (or longer if there are symptoms suggestive of hypoglycaemia or ongoing concerns with BGLs).
- Refer to Management of Hypoglycaemia on the Ketogenic Diet flowchart (Appendix 1) for details of hypoglycaemia management (also available on www.pennsw.com.au).
- Epilepsy CNC to discuss the most appropriate and preferred method of BGL monitoring, for example, by parents, Hospital in the Home (HITH) or GP, considering patient's age, carbohydrate intake prior to ketogenic diet initiation, and parents' capability to monitor BGLs in the home setting.
- Medical staff to refer to HITH services if required.
- If monitoring is to be done by parents, Epilepsy CNC will provide training on how to use glucometer. The blood monitoring units may be loaned during transition onto the diet.(Test stix must be purchased by the family). Education will be given on when to seek medical attention (e.g. if low BGLs persist and do not respond to emergency plan or if patient has new onset seizures which could be related to low BGLs)
- Parent/carer to notify Epilepsy CNC and dietitian of BGL readings via email or phone during early transition onto the diet.
- Parents to contact the Neurology fellow via hospital switch or present to the Emergency Department if BGLs remain low or the child remains symptomatic of hypoglycaemia.

Monitoring ketone levels

- Method of ketone testing is determined on a case by case basis. Epilepsy CNC to discuss this with parents/carers.
- Urine ketones to be tested initially with every urine output until ketosis is established, then twice a day until urine ketone levels are stable
- Recommend testing first thing in the morning and after school
- Blood betahydroxybutyrate (blood ketone) to be checked when full MAKD is fully established and child is in ketosis

Appointment at Ambulatory Care Unit

- Organise a follow-up appointment one month after initiation of the diet.

Follow-up

First month review and subsequent 3 – 6 monthly reviews, trouble-shooting and further education provision

Location: Ambulatory Care Unit / Outpatients

Aim: To provide further education and counselling to assist with compliance on modified Atkins ketogenic diet

Tasks	Responsibilities
☐ Document anthropometry parameters (weight, height or length, and head circumference if appropriate) ☐ Assess efficacy and tolerance of the diet ☐ Check compliance with the ketogenic diet ☐ Assess side effects and discuss management of side effects ☐ Assess risk of gastrointestinal intolerance and micro aspiration ☐ Blood tests including blood ketones, blood gas and electrolytes ☐ Urine test including calcium:creatinine and citrate:creatinine ratio ☐ Issue PBS scripts if required ☐ Ensure subsequent follow-up with patient's own Neurologist ☐ Check that an "allergy" for "ketogenic diet" is in Powerchart for new patients.	Medical staff
☐ Monitoring of ketone testing and BGL testing (if parents are doing self BGL monitoring in the home setting) ☐ Assess motivation of patient, parents/carers to remain on the diet ☐ Trouble shooting ☐ Arrange routine blood screening (refer to Appendix 2) ☐ Ongoing assistance with preparation for routine procedures and admissions post initiation of the MAKD	Epilepsy CNC
☐ Conduct full nutrition assessment, including anthropometry parameters (weight, height, BMI) ☐ Review appropriateness of diet (energy, fluid, macronutrients) ☐ Review blood sugars and urine ketones and adjust diet accordingly ☐ Review side effects and identify appropriate treatment ☐ Review supplementation (multivitamin and Calcium & Vitamin D) ☐ Assess nutrition knowledge of parents/carers ☐ Assess compliance to therapy – food provision & preparation, economic considerations ☐ Assess motivation of patient, parents/carers to remain on the diet ☐ Provide further dietary education and support if required ☐ Consider fine-tuning of the diet or supplements to assist in enhancing ketosis	Dietitian
☐ Review medications and confirm carbohydrate content in medications	Pharmacist

Essential information/resources to give to parents

Ensure family has written information in a pack including the following:

- Emergency plan/letter
- Trouble shooting information sheet
- Written dietary information
- PBS scripts
- Research programs running through the ketogenic diet program

3 References

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Appendix 1 Management of Hypoglycaemia on the Ketogenic Diet flowchart

Also available on SCHN Intranet and http://www.pennsw.com.au

Appendix 2 Recipe for Potassium Citrate Mixture (sugar free)

Once ketosis is reached (blood betahydroxybutyrate >1mmol/ml) the patient will need supplementation with sugar free potassium citrate mixture in order to prevent nephrocalcinosis and hypocitraturia which predisposes to renal calculi whilst on the ketogenic diet.

This mixture is usually provided as an oral liquid prepared by the SCHN pharmacy for the family.

The dose is 1.9mmol/kg/day in 2-3 divided doses.

An in-hospital script will need to be written to provide the family with the Potassium citrate Mixture (sugar free).

Sometimes families will request a script for the local community pharmacy particularly for patients living in regional NSW.

An Authority PBS script for extra-contemporaneous supply of an oral mixture is required. This can be done by ringing PBS on 1800888333 and asking to speak to the PBS pharmacist

Product: Potassium Citrate Mixture (**sugar free**) – 1.9 mmol/ml
Strength (Each mL contains 200mg potassium citrate, 40mg citric acid monohydrate)
Shelf life: 30 days
Storage Conditions: Room temperature (less than 25 ^{o}C)
Recipe:

Ingredient	Unit Qty
Potassium Citrate (note: potassium citrate BP=potassium citrate monohydrate)	20g
Citric acid monohydrate	4g
Methyl hydroxybenzoate 5% solution	1mL
Water for irrigation	To 100mL

Formulae is from Australian Pharmaceutical Formulary (APF) 21st edition with syrup removed

The team also occasionally uses Urocit-K® tablets which contains 10mEq (~10mmol) Potassium Citrate (or 1.08 g potassium citrate) per tablet - https://www.nps.org.au/medicine-finder/urocit-k-tablets

There is a registered proprietary potassium citrate mixture called Uricosal® (contains 67% sucrose) which is not to be used in patients on ketogenic diet.

Appendix 3: Long Term Monitoring of the Ketogenic Diet

Bloods	**3 M**	**6 M**	**9 M**	**1 Year**	**15m**	**18m**
Blood Ketone (betahydroxybutyrate)	☐	☐	☐	☐	☐	☐
Full blood count	☐	☐	☐	☐	☐	☐
Electrolytes (Sodium, potassium, bicarbonate, urea, creatinine), Calcium, Magnesium, Phosphate)	☐	☐	☐	☐	☐	☐
Serum liver profile (Total Protein Albumin, AST, ALT)	☐	☐	☐	☐	☐	☐
Lipid profile (triglycerides and cholesterol)	☐	☐	☐	☐	☐	☐
Plasma carnitines (total, free, acyl) (optional –available at SCH)	☐	☐	☐	☐	☐	☐
Glucose, lactate	☐	☐	☐	☐	☐	☐
Plasma amino acids – to assess for protein and amino acid deficiency	☐	☐	☐	☐	☐	☐
Serum kidney profile (Sodium, potassium, blood urea nitrogen, creatinine)	☐	☐	☐	☐	☐	☐
Urine Calcium:Creatinine ratio	☐	☐	☐	☐	☐	☐
Urine Citrate:Creatinine ratio	☐	☐	☐	☐	☐	☐
Micronutrients						
25 OH Vitamin D and Parathyroid hormone	☐	☐	☐	☐	☐	☐
Zinc	☐	☐	☐	☐	☐	☐
Selenium	☐	☐	☐	☐	☐	☐
Active B12	☐	☐	☐	☐	☐	☐
Total Vitamin B 12	☐	☐	☐	☐	☐	☐
Folate	☐	☐	☐	☐	☐	☐
Iron Studies	☐	☐	☐	☐	☐	☐
Plasma Amino Acids (optional –available at SCH)	☐	☐	☐	☐	☐	☐
Other						
Check for symptoms of vitamin C deficiency	☐	☐	☐	☐	☐	☐

Medical Tests	**3 M**	**6 M**	**9 M**	**1 Year**	**18 M**	**2 Years**
ECG at 3m						
Renal and abdominal Ultrasound if nephrocalcinosis						
DEXA- Bones – bone age/bone density (note: more useful for children > 4 years)						

Urine metabolic screens, plasma amino acids and carnitine profiles are not available at outside pathology centres and may cost the patient a considerable amount. SEALS has agreed to pay for Plasma amino acids and carnitine profiles provided a SEALS pathology form is completed with ketogenic diet written in the clinical section of the form and blood is collected at a SEALS pathology centre (Randwick, St George, Wollongong Hospital, and Sutherland Hospital only). Blood collected at other centres – the patient will be charged and will be non-medicare rebatable. Please note that CHW will not perform these metabolic tests unless the clinical indication (ie Patient on ketogenic diet) is completed.

**** Patients on a ketogenic diet need special precautions when they are unwell or when they are fasting due to the risk of metabolic acidosis, hypoglycaemia and dehydration.**

Appendix 4: What to do if a sick ketogenic diet patient presents to Emergency Department

If a patient on the Ketogenic Diet presents to your Emergency Department due to illness, the Ketogenic Diet should take second place to the treatment necessary for the presenting complaint. However, wherever possible, sugar-free medications (or those with minimal carbohydrate content) should be used and the treating team should aim to maintain ketosis to avoid seizure activity, and avoid hypoglycaemia.

Refer to the patient's Emergency Letter (if available) and consider the following recommendations specific to Ketogenic Diet patients:

Resuscitation:

- Resuscitate as per usual protocols
- Administer normal saline bolus (as per child's age and requirements)

Investigations specific to being on the diet:

- Unwell KD patients require close and regular monitoring
- Avoid hypoglycaemia, particularly if vomiting or diarrhoea present (at least 4-hourly bedside checks of **blood glucose** required) (Refer to: Management of Hypoglycaemia on the Ketogenic Diet flowchart for more detailed information: www.pennsw.com.au).
- Avoid high ketone levels (at least 4-hourly urine samples to check ketone levels). If the urine ketone levels are high perform blood ketone (betahydroxybutyrate). Avoid blood ketone level of > 7 mmol/L.
- Venous gas, lactate and glucose (to check for metabolic and lactic acidosis)
- Electrolytes (look for hypernatraemia, hyper/hypokalaemia)
- Blood glucose (to look for hypoglycaemia)
- Blood lactate (looking for elevated lactate)

Hydration:

- Where hydration is required, aim to use fluids containing the lowest possible glucose concentration, while maintaining blood glucose levels in the normal range.
- Oral hydration: offer clear fluids low in carbohydrate (e.g. hydrolyte, gastrolyte, diet cordial). Dilute rehydration fluids 2:1 if necessary to avoid loss of ketosis.
- IV hydration: 0.9% sodium chloride as IV fluid in most cases but if there are concerns re: hypoglycaemia or prolonged IV use, consider adding **low** glucose content fluids to IV (e.g. 2.5%glucose)
- NG hydration: if there are concerns re: hypoglycaemia, consider using gastrolyte® as this contains no more than 3% glucose
- **Medical staff should chart glucose, glucagon or polyjoule as necessary.**

Medications:

- **It is essential that any newly prescribed medications are sugar-free** (or contain minimal carbohydrate content)**. This applies to all medication including cough syrups, pain relief and antibiotics (e.g. syrups may need to be changed to sugar-free liquids or to crushable tablets)**. Compounding can be discussed with the patient's local Pharmacy.
- Consider placing an alert of avoiding sugar and carbohydrates in the Allergies section of the medication chart for patients on the ketogenic diet

If additional advice or support is required, contact the patient's treating Ketogenic team or the relevant Neurology Fellow on-call directly.

Appendix 5: What to do if a ketogenic diet patient presents for a routine procedure

If a patient on the Ketogenic Diet requires a routine surgical or medical procedure, it is important to follow the recommendations below:

Fasting and BGL's:

- Fasting poses a significant risk for Ketogenic Diet patients
- Try to avoid unnecessary fasting prior to procedures
- If BGL's drop while fasting, consider using IV 0.9% sodium chloride + 2.5% glucose to achieve normal glucose levels (Refer to: Management of Hypoglycaemia on the Ketogenic Diet flowchart for more detailed information: www.pennsw.com.au).
- The anaesthetist should be informed regarding regular monitoring of BGL's intra-operatively
- BGL's should be performed every 60 minutes

Operative lists:

- Ketogenic Diet patients should be treated as you would treat a diabetic patient i.e. they should be **placed FIRST or early on the operative list**

IV Fluids:

- The preferred fluid is 0.9% sodium chloride (except where hypoglycaemia present, see above)

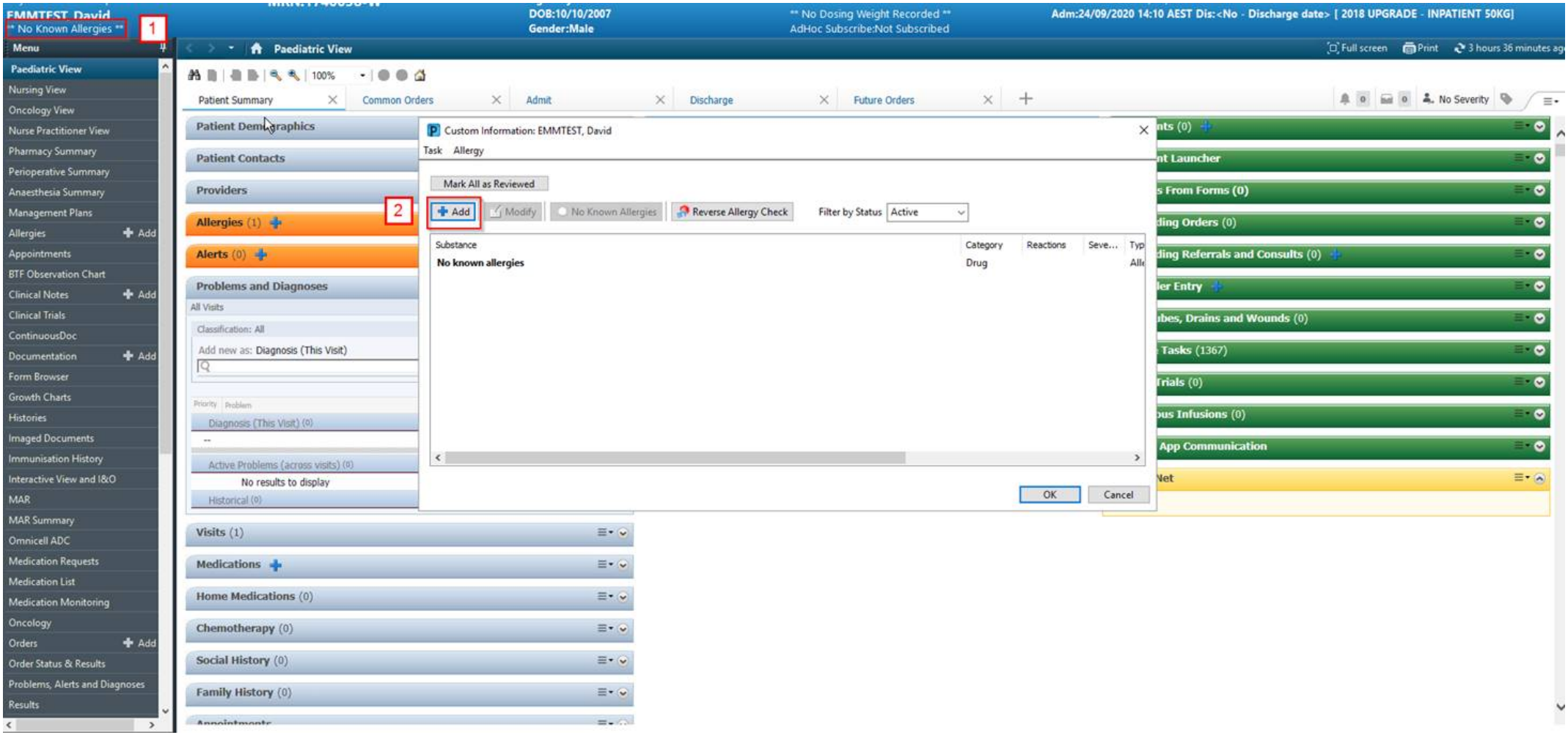
Following the procedure/cessation of fasting period:

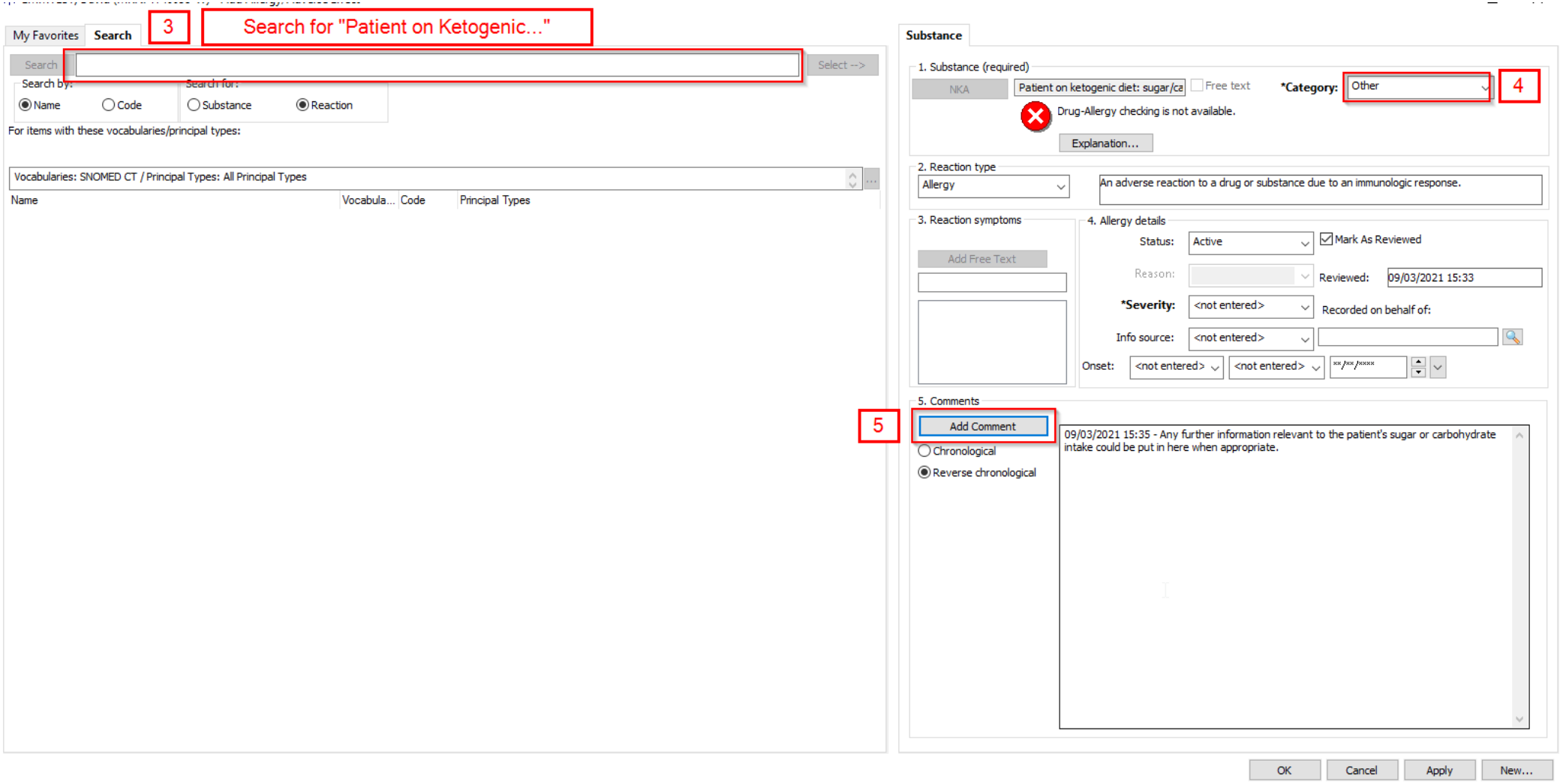
- Reinstitute the Ketogenic Diet once fasting/procedure is over
- Consider using the Ketocal formula NG at 4:1 or 3:1 if the patient cannot yet resume their usual ketogenic diet (e.g. Ortho. and ENT procedures).

Medications:

It is essential that any newly prescribed medications are sugar-free (or contain minimal carbohydrate content). This applies to all medications including pain relief (paracetamol and ibuprofen syrups have large quantities of sugar) and antibiotics (e.g. syrups may need to be changed to sugar-free liquid or use crushable tablets) Information can be obtained from hospital pharmacy or by calling the drug companies via their medicines information department.

Appendix 6: eMM - Allergies




3
Search for "Patient on Ketogenic..."
My Favorites
Search
Search
Select -->
Search by:
Name
Code
Search for:
Substance
Reaction
For items with these vocabularies/principal types:
Vocabularies: SNOMED CT / Principal Types: All Principal Types
Name
Vocabula...
Code
Principal Types
Substance
1. Substance (required)
NKA
Patient on ketogenic diet: sugar/ca
Free text
*Category:
Other
4
Drug-Allergy checking is not available.
Explanation...
2. Reaction type
Allergy
An adverse reaction to a drug or substance due to an immunologic response.
3. Reaction symptoms
Add Free Text
4. Allergy details
Status:
Active
Mark As Reviewed
Reason:
Reviewed:
09/03/2021 15:33
*Severity:
<not entered>
Recorded on behalf of:
Info source:
<not entered>
Onset:
<not entered>
<not entered>
//****
5. Comments
5
Add Comment
Chronological
Reverse chronological
09/03/2021 15:35 - Any further information relevant to the patient's sugar or carbohydrate intake could be put in here when appropriate.
OK
Cancel
Apply
New...