

48 HOUR URINE COLLECTION A DIAGNOSTIC TEST FOR WILSONS DISEASE GUIDELINE		
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Approved By Name: Fionnuala O'Neill Title: Nurse Practice Coordinator	Signature: Date: April 2018 Ironnwala Divell	
Authorised By Name: Rachel Kenna Title: Director of Nursing	Signature: Date: April 2018	
Author/s	Name: Geraldine O'Shea Title: Clinical Nurse Specialist, GI Unit	
Location of Copies	On Hospital Intranet and locally in department	

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Date of Issue: April 2018	Page 2 of 6	Croman

CONTENTS

Page Number

1.0	Introduction	3
2.0	Definition of Guidelines	3
3.0	Applicable to	3
4.0	Objectives of the Guideline	3
5.0	References (as necessary)	4

Our Lady's Children's Hospital, Crumlin		Ospidéal Mhuire na Leanaí.
Document Name: 48 Hour Urine Collection - A Diagnostic Test For Wilsons Disease Guideline		Cromghlinn
Reference Number: 48HUCDTWD-01-2018-GOS-V1 Version Number: V1		Our Lady's Children's Hospital, Crumlin
Date of Issue: April 2018	Page 3 of 6	Cromun

1.0 Introduction

This is a test that is carried out in all children who are being investigated for liver disease to out rule Wilson's Disease.

Wilson's Disease is a rare condition with an incidence of 30:1,000000 (Fryman 1990). It is a genetic disorder caused by absence or decreased function of ATP 7B gene. This gene encodes a metal transporting P-type adenosine, triphopsphatase (ATPase) which is expressed mainly in the hepatocytes and functions in the transport of copper within the hepatocytes. Absent or decreased function of ATP7B protein leads to decreased hepatocellular excretion of copper into bile. This results in hepatic copper accumulation and injury. Eventually copper is released into the bloodstream and deposited in other organs notably the brain, kidneys and cornea

2.0 Definition of Guidelines

The 24 urinary copper excretion after penicillamine challenge is one of the diagnostic tools used in determining a diagnosis of Wilson's Disease.

It is the most accurate diagnostic test in Wilson's Disease (Martins da Costa et al 1992).

This test involves a 48 hour urine collection which incorporates the 1st 24hours urinary copper collection) followed by the 2nd 24hours urine collection in conjunction with the administration of penicillamine in two divided doses, 12 hours apart).

3.0 Applicable to

Nursing staff caring for a child for whom this investigation has been requested by the medical team.

4.0 Objectives of Guidelines

To ensure urine collection is completed accurately. All urine must be collected in the 48 hour period. If any urine is missed or discarded the test is invalid and the collection process must begin again

ACTION	RATIONALE
Explain procedure to child and parent and the duration of the 48 hour urine collection.	Proper preparation may decrease a child's anxiety and encourage co-operation (Ball and Binder 2008)
Explain that Penicillamine will be administered as prescribed after the 1 st 24 hour urine collection is completed and administered again 12 hours later	Penicillamine solubilises copper and allows the stores to be excreted in urine.
Nurse to phone the Biochemistry laboratory to inform them that containers are required for 48 hour urine collection	To allow the Laboratory staff time to prepare the acid washed containers needed for the collection

Our Lady's Children's Hospital, Crumlin		Ospidéal Mhuire na Leanaí.
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Reference Number: 48HUCDTWD-01-2018-GOS-V1 Version Number: V1		Our Lady's Children's Hospital, Crumin
Date of Issue: April 2018	Page 4 of 6	crumun

Nurse to ensure team have prescribed Penicillamine	To ensure that drug is available at the specific time required so test can be completed successfully.
Dose: <8 yrs 250mg	To ensure dosage is correct as per drug formulary/pharmacy guidelines.
>8yrs 500mg given 12 hours apart at start of 2 nd 24 hour collection and 12 hours after the initial dose	
Ensure that there is a collection jug available on the ward/unit for sole use by the child for 48 hour period.	To ensure collection of all urine. To avoid cross contamination as per IPCC OLCHC.
Collect containers from the Biochemistry Laboratory. (4 containers) 2 containers marked '1' and 2 containers marked '2'	2 containers should be provided for each 24hour period in case more urine than can be contained in one is passed. Container marked 'No. 1' will be used for the 1 st 24hours and container marked No. 2 will be used for the 2 nd 24 hours
Record the patients urinary output in the Intake and Output Record Sheet	Use to refer for start and finish times and administration of penicillamine
First 2	4 hours
First 2 Discard the 1 st urine of the day. Ask the child to void into the toilet as normal.	This is the start time of the first 24 hour collection.
Discard the 1 st urine of the day. Ask the child to	
Discard the 1 st urine of the day. Ask the child to void into the toilet as normal. Record the start time on the container marked No 1 and on the output record	
Discard the 1 st urine of the day. Ask the child to void into the toilet as normal. Record the start time on the container marked No 1 and on the output record Continue to pass all urine for the next 24 hours	This is the start time of the first 24 hour collection. If all urine is not collected the test is invalid and/or could result in a false negative or false positive
Discard the 1 st urine of the day. Ask the child to void into the toilet as normal. Record the start time on the container marked No 1 and on the output record Continue to pass all urine for the next 24 hours into a jug and empty into container marked No. 1 When 24 hours of collection from start time is completed, record the time on container 1 and the	This is the start time of the first 24 hour collection. If all urine is not collected the test is invalid and/or could result in a false negative or false positive (Pandit et al 2002) This is the end of the 1st 24 hour urine collection. Store container in a safe area (i.e. the sluice room and ensure it is not discarded) As per An Bord Altranais Guidance to Nurses and midwives on medication management (2007)
Discard the 1 st urine of the day. Ask the child to void into the toilet as normal. Record the start time on the container marked No 1 and on the output record Continue to pass all urine for the next 24 hours into a jug and empty into container marked No. 1 When 24 hours of collection from start time is completed, record the time on container 1 and the Intake and Output Record Sheet Ensure child has identity band insitu and medication is checked appropriately. Administer	This is the start time of the first 24 hour collection. If all urine is not collected the test is invalid and/or could result in a false negative or false positive (Pandit et al 2002) This is the end of the 1st 24 hour urine collection. Store container in a safe area (i.e. the sluice room and ensure it is not discarded) As per An Bord Altranais Guidance to Nurses and

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Reference Number: 48HUCDTWD-01-2018-GOS-V1 Version Number: V1		Our Lady's Children's Hospital, Crumlin
Date of Issue: April 2018	Page 5 of 6	crumun

24 hours in passed into a jug and decanted into Container marked no 2	and will have to be undertaken at a later date.	
Administer the second dose of Penicillamine 12 hours after first dose is administered.	If the 2 nd dose of Penicillamine is not given this test is null and void	
Record on container no 2 and Intake and Output Record Sheet when the second dose of Penicillamine is administered	To ensure validity of the test	
Collect all urine for the next 24 hours and empty into container marked No 2.	Roberts and Schilsky (2006) state the amount of copper excreted in urine in a 24 hour period may be useful in diagnosing Wilsons Disease and for monitoring treatment	
Record the completion time on container marked No 2 and Intake and Output Record Sheet	This completes the 48 hour urine collection	
Containers to be brought to the Biochemistry laboratory when the test is completed.	To ensure prompt delivery, in order for the laboratory to complete sample testing.	

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Reference Number: 48HUCDTWD-01-2018-GOS-V1 Version Number: V1		Our Lady's Children's Hospital, Crumlin
Date of Issue: April 2018	Page 6 of 6	crumun

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