

STANDARD OPERATING PROCEDURE (SOP) W & C Division

Directorate	Paediatric Directorate
Ward/Service	Cystic Fibrosis Service
Title of SOP	Management of care of Infants diagnosed with Cystic Fibrosis through Neonatal Screening
SOP Number	2
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OBJECTIVES	<p>This SOP will describe the process for managing the care of children and their families during the first year following a diagnosis of CF through Neonatal screening.</p> <p>To promote high quality of care and ensure patient information is consistent with CF Trust guidelines</p>	
SCOPE	<p>This SOP applies to patients diagnosed with Cystic Fibrosis through Neonatal screening programme</p>	
TARGET GROUP	<p>This SOP applies to patients with a diagnosis with Cystic Fibrosis attending the Teesside Cystic Fibrosis Service</p>	
EVIDENCE TO SUPPORT PROCEDURE	<p>Standards for the Clinical care of Children and Adults with Cystic Fibrosis in the UK CF Trust , December 2011</p>	
CONTENTS	<p>Responsibilities</p> <p>Definitions and Abbreviations</p> <p>Informing the family</p> <p>Preparation for Consultation</p> <p>Diagnosis Consultation</p> <p>1st Home visit</p> <p>Week 2</p> <p>1 Monthly review</p> <p>6 Monthly review</p> <p>1st year review</p>	<p>2</p> <p>2</p> <p>2</p> <p>3</p> <p>3</p> <p>4</p> <p>4</p> <p>5</p> <p>5</p> <p>5</p>

Responsibilities

The following persons have responsibilities within the SOP
 Paediatric Consultant with responsibility for Children with CF
 Specialist Nurse for Cystic Fibrosis
 Clinical Specialist Physiotherapist
 Senior Clinical Specialist Dietician
 Specialist Pharmacist

Definitions and Abbreviations

Abbreviation	Definition
SOP	Standard operating procedure
CF	Cystic Fibrosis
HV	Health Visitor
SpN	Specialist Nurse
POPD	Children's Out Patient Department
RSL	Regional Screening Laboratory

	Sequence of Clinical Procedure	Rationale/Additional Information	Lead
1.0	Informing the family		
1.1	Teesside CF Team are informed of a potential new patient by the CF Team at RVI		SpN
1.2	Baby's name, Address, DOB. GP, Mothers name, Genotype (If known) NHS number recorded on new diagnosis record sheet	To ensure correct identification of baby	SpN
1.3	SpN contacts the Area lead HV and informs them of the date and time of appointment.		SpN
1.4	Lead HV and Family HV visit the family to give information regarding the screening result and an appointment date and time to see the CF team at hospital within 24 hours	HV will not visit on a Thursday or Friday to minimise Parental stress over a weekend when no support is available	HV
1.5	SpN offers support and advice to Lead HV if required via telephone.		SpN

2.0	Preparation for Consultation		
2.1	Register the baby onto Camis system		SpN
2.2	Prepare Notes		SpN
2.3	Book Clinic room		SpN
2.4	Organise prescription		SpN
3.0	Diagnosis Consultation		
3.1	Parents and Baby attend POPD and are shown into a consultation room. Extended family members are welcome as appropriate to support parents		SpN
3.2	The baby's length, weight and head circumference are measured and plotted on the centile chart	To establish baseline measurements	SpN
3.3	Family are introduced to Paediatric Consultant and SpN initially and the diagnosis of CF is explained.	This is a very distressing time for the family and the CF team allow lots of time for the family to understand the information and ask questions	Consultant
3.4	Sweat Test – if 2 genes identified then wait until 6 month review	To confirm Diagnosis Within 24hrs of consultation	
3.5	Physiotherapist and Dietician are then introduced to the family.	Dependent on presentation of baby	Consultant
3.6	Information leaflet is given on how to give Pancreatic enzyme replacement therapy		Dietitian
3.7	If the baby is clinically well there is no need for admission*	The following conditions may require admission to Paediatric unit: Failure to regain birth weight Clinical signs of Respiratory infection Social history raising safeguarding or coping concerns	Consultant

3.8	The parents are given the following information:- CF Trust – The Facts CF Trust Website Contact Numbers On-call service	To ensure evidence based information is available	SpN
3.9	An appointment is given for the SpN to visit the family at home within 2 working days.		SpN
4.0	1st Home visit by SpN		
4.1	SpN will review the information given at the diagnosis consultation and ascertain how much information the parents have retained	Answer questions openly and honestly	SpN
4.2	Introduce Genetics service and cascade screening		SpN
4.3	Stool specimen for Faecal elastase	To establish Pancreatic status	SpN
4.4	Urine specimen for electrolytes	To identify if Sodium supplements are required	SpN
4.5	Organise Sweat test appointment for siblings	To confirm Diagnosis	SpN
4.6	Organise repeat sweat test for patient if initial test insufficient		SpN
4.7	Give appointment to attend CF Clinic		SpN
5.0	Week 2		
5.1	Home visit by SpN and physiotherapist		
5.2	Commence prophylactic antibiotics and Vitamin supplements unless clinically indicated earlier		SpN
5.3	Introduce physiotherapy techniques		Physiotherapist
5.4	Obtain Consent for PORT CF		SpN

6.0	1st Monthly Review		
6.1	To attend CF Clinic every month for 1 st year	To monitor clinical condition and allow early intervention	Team
6.2	October – March to have Palivizumab vaccinations	To minimise risk of RSV infection	Consultant
6.3	If over 6 months age September to November – to have Flu Vaccination	To minimise risk of infection	Consultant
7.0	6 Months Review		
7.1	Blood sample for Vitamin A, D and E	To monitor supplementation	SpN
7.2	Repeat stool elastase if originally pancreatic sufficient	To confirm Pancreatic status	SpN
7.3	Review Knowledge of CF		Team
7.4	Re discuss Genetics service and referral process		SpN
8.0	1 Year Review		
8.1	Annual review	To monitor clinical status	Team
8.2	Reduce clinic appointments to every 8 weeks unless clinically indicated		Team

Developed By:	AUTHOR TITLE (NAME)	JOB TITLE
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	Teesside Cystic Fibrosis Team	October 2014