# THE MANAGEMENT OF NEW DIAGNOSIS CF/CRMS PATIENTS AT OUR CF CENTER

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PEDIATRIC CF CLINIC PROGRAM COORDINATOR

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## CF CLINIC WORKFLOW
## CATEGORY B VS CATEGORY C

- MMP CF Clinic will be notified by fax from the Maine NBS program.
- Our colleagues in Genetics will communicate with the PCP and arrange a sweat test with genetic counseling
- Pts with indeterminate/borderline sweat chloride values (30-59 mmols/L) would be referred to the CF Center for further evaluation.
- If a pt has 2 borderline sweat chloride tests with one CF mutation they are followed by our clinic as CRMS.
- CF Center will be notified by fax and phone from the Maine NBS program.
- CF Provider will be informed by clinic staff and will reach out to the PCP to arrange for an urgent Pulmonary consult.
- Newborn screening results will be given to the family by the PCP.
- CF Program Coordinator will facilitate that first visit in our clinic within 48 hours of the family receiving those results where they will meet with one of our CF Providers and a Genetic Counselor.
- The CF clinic will arrange confirmatory sweat testing when the baby is >2 weeks old AND 2kg

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## CF clinic follow up for pts with CRMS

Cystic Fibrosis transmembrane conductance regulator-related metabolic syndrome

The CFF developed Clinical Care Guidelines that recommended that pts be seen by a CF specialist and have a repeat sweat test by 2 months of age. If the sweat chloride remains borderline they have a third sweat test at 6 months of age. If they remain borderline and symptom free we will see them twice a year for the first year of life and annually thereafter. We will order a CMP, CBC, Vitamin A, D & E levels, GGT and INR , 2 view CXR and CF gag culture annually.

These patients are at increased risk for the development of CF-like symptoms. Evolving signs and symptoms, new information about disease causing mutations or change in sweat chloride concentrations may lead to a diagnosis of CF.

We do not routinely start therapies, eg salt replacement, vitamins, airway clearance unless clinically indicated.

Borowitz et al (J Pediatrics 2009)

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## CF Clinic Follow up for patients with CF

Newly diagnosed CF patients are seen in our clinic within 48 hours of the family being notified of the positive newborn screen.

At this visit the CF Provider and Genetic Counselor will discuss the diagnosis, do a physical exam of the infant, provide support and answer any questions the family may have.
We will collect baseline vitals including weight and height, arrange to collect a stool sample for Fecal Elastase, perform a gag throat culture and will likely start salt and enzyme replacement.

We may involve our CF LCSW at this visit to attend to any urgent needs for additional support the family may have. The families are given the contact numbers for the clinic, our nurse triage line, the LCSW and the RN Coordinator along with our “New Patient Packet” before they leave this appointment, the contents of which are shown in the following slide.

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## RESOURCES IN OUR NEW PATIENT PACKET

The CFF Introduction to CF for pts and families

Photos and bios of our CF Team members

Pediatric CF Clinic brochure

Our CF center's "Pathway For Infant Nutrition"


"Navigating the world of CF" Helpful resources and links

Contact information for our local CFF NNE Chapter

CFF Community Programs

Clinic contact details

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## CF Clinic follow up for patients with CF continued

We ask the family to return to clinic within 1 month or sooner if they have concerns. We will often combine this appointment with a sweat test at the lab at MMC.
At this visit they will be able to meet the CF Providers and the other members of our Multidisciplinary Team including PT, RT, Research Coordinator, CF Pharmacist, Nutritionist, LCSW and RN Coordinator.

We see these babies monthly for the first year of life and quarterly thereafter.
We also work closely with our GI , ENDO and ENT providers and may ask our PCP colleagues to place referrals to involve these specialists in the patient's care.

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We value our partnership with Primary Care Physicians. Our office visit notes will be routed to the PCP on file and we welcome any and all communication that helps us participate in their care.

Please contact ann.ladner@mainehealth.org (207) 662 5719 with requests for any additional information

THANK YOU FOR LISTENING TO OUR PRESENTATION.

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