

## NSC-NL Passes Reaccreditation

To ensure that it maintains quality standards, NSC-Northern Luzon (NSC-NL) underwent and successfully earned its reaccreditation status for another three years.

The accreditation, which was led by a team from DOH and Newborn Screening Reference Center (NSRC) with some international newborn screening experts, was conducted on July 26-27, 2019, at the Mariano Marcos Memorial Hospital and Medical Center (MMHMC), Batac City, Ilocos Norte.

"The commitment and dedication of NSC-NL staff and management are to be commended," said Dr. Bradford Therrell, Jr., member of the international experts during the reaccreditation. "The staff is young, enthusiastic, excited, and well-grounded about the program. NSC-NL maintains a good relationship with the host organization. The laboratory and non-laboratory operations were reviewed and appear to be in a stable state."

The NSRC Accreditation Team was composed of Dr. Therrell, Director of the National Newborn Screening and Global Resource Center in Texas, USA; Dr. Veronica Wiley, Director of the New South Wales Newborn Screening Program in Australia; and Dr. Florencio Dizon, Quality Assurance Consultant from the NSRC of the Philippines. Ma. Elouisa Reyes, Margarita Aziza Canlas, and Ma. Truda Escoreal from NSRC were also in attendance to assist the team.

Also present were Dr. Jose B. Orosa III, Chief of Medical Professional Staff of MMHMC; Cyrus Jed Ramos, NBS Program Manager of DOH-CHD Region 1; Dr. Annabelle Paguirigan, NBS Medical Coordinator of DOH-CHD Region 2; and Dr. Ma. Paz Virginia K. Otayza and all of NSC-NL staff.

The accreditation process consisted of an opening meeting, data and accomplishment presentation by the center, parallel test, checking of facility, workflow observation, and checking of documents and competencies as required by the Philippine PEAS tool and in compliance with ISO standards. The activity culminated in the recommendations after the external audits.

NSC-NL started offering NBS services to Ilocos and Cagayan Valley regions in 2017. It is hosted by MMHMC and is currently headed by Dr. Maria Paz Virginia Otayza (Unit Head), Dr. Modesty Leaño (Laboratory Manager), Dr. Karla Modesta Albano (Follow-Up Head), and Anthony James Almazan (Program Manager). **AJAlmazan**



Top: Dr. Bradford Therrell (leftmost) conducts an interview with the Follow-Up Team. Center: Dr. Veronica Wiley (second from left) checks the process flow of the laboratory with Ma. Truda Escoreal (leftmost). Bottom: Dr. Florencio Dizon (rightmost) observes the process of Tandem Mass Spectrometry (MS/MS).

## PHO Benguet Conducts Common Metabolic Disorders Forum



Benguet Governor Melchor Diclas (seated second from right) joins the organizers and participants during the conduct of the Metabolic Disorders Forum held on August 23 in La Trinidad, Benguet.

The Provincial Health Office (PHO) of Benguet, in partnership with Baguio General Hospital and Medical Center, conducted a forum on common metabolic disorders at the PHO Conference Hall, La Trinidad, Benguet, on August 23, 2019.

Governor Melchor Diclas attended the event and addressed more than 40 doctors, nurses, medical technologists, and midwives from different hospitals and health centers in Benguet. The governor declared his support to newborn screening and other health programs in the province.

The activity was made possible through the efforts of Dr. Nora Ruiz, PHO Head, and Dr. Antonette Agpas, Maternal, Newborn, and Child Health and Nutrition (MNCHN) Head, together with Rosana Tabangcura, provincial NBS coordinator.

The forum aimed to orient health workers in the long-term management of common metabolic disorders identified through newborn screening test. Dr. Sharon Gawigawen, pediatric endocrinologist, discussed common metabolic disorders and

their management, while Florenz Roniello Nastor, long-term follow-up nurse, talked about follow-up care and monitoring of children with these disorders. Meanwhile, Dominga Dawe, nutritionist/dietitian at the Baguio General Hospital and Medical Center, gave a lecture on nutritional requirements for patients.

The three resource persons are part of the NBSCC Team who is in charge of the long-term follow-up management and monitoring of NBS confirmed patients in and around the Cordillera region.

The participants, for their part, vowed to help in the promotion and strengthening of the NBS Program in the province and region.

With the success of the forum, Benguet PHO committed to hold another forum in the future to cover all of the newborn screening facilities in Benguet. It also pledged to help in locating patients in the province who were lost during the follow-up stages. **FRNastor**

## IPHO-Maguindanao Hold 2019 Semi-Annual PIR

The Integrated Provincial Health Office (IPHO)-Maguindanao held its Semi-Annual Program Implementation Review (PIR) and Catch-up Planning Workshop at Greenleaf Hotel, General Santos City, on July 3-6, 2019.

The event was attended by health workers and heads in Maguindanao Province as well as representatives from health partners such as the University Research Company, UNFPA, Pilipinas Shell Foundation, Inc., ARCHES Project, and the Ministry of Health of the Bangsamoro Autonomous Region in Muslim Mindanao.



Left to right: Kim Ian Tiu (Partner from UNFPA), Dr. Elizabeth Samama (Chief of Hospital at Buluan District Hosp, now OIC Provincial Health Officer at Maguindanao Province), Anisa Matuan (Planning Officer-Ministry of Health BARM), Dr. Tahir B. Sulaik (Provincial Health Officer II at Maguindanao Province), Dr. Ma. Melissa T. Miranda (Partner from University Research Company or URC), Dr. Moh. Ariff Baguidal (PHO I at Mag. Province), Dr. Geraldine A. Macapeges (Chief at Technical Division IPHO Mag), and partner from Pilipinas Shell Foundation, Inc.

Continued on page 8 . . .



DOH-CHD Northern Mindanao Regional Nurse Coordinator Angel Balansag (second row, seated second from left) and NSC-M Unit-Head Dr. Conchita G. Abarquez pose for a photo with the participants from Lanao del Norte during the culmination of the training.

## Mindanao CHDs, NSC-M Hold NBS Trainings

As partner agency of the ENBS Program, the CHDs, in coordination with NSC-M, see to it that with the transition to ENBS, all facility coordinators are equipped with the proper knowledge and skills in implementing the program.

Two NBS training programs conducted in August 2019 were the result of this commitment.

On August 1-2, 2019, through the efforts of the PHO of Lanao del Norte and in coordination with DOH-CHD Northern Mindanao, 26 participants from all the municipalities of the province attended the training at Pearlmont Inn, Cagayan de Oro City. The hands-on training on heel prick was conducted at the Northern Mindanao Medical Center. The activity was

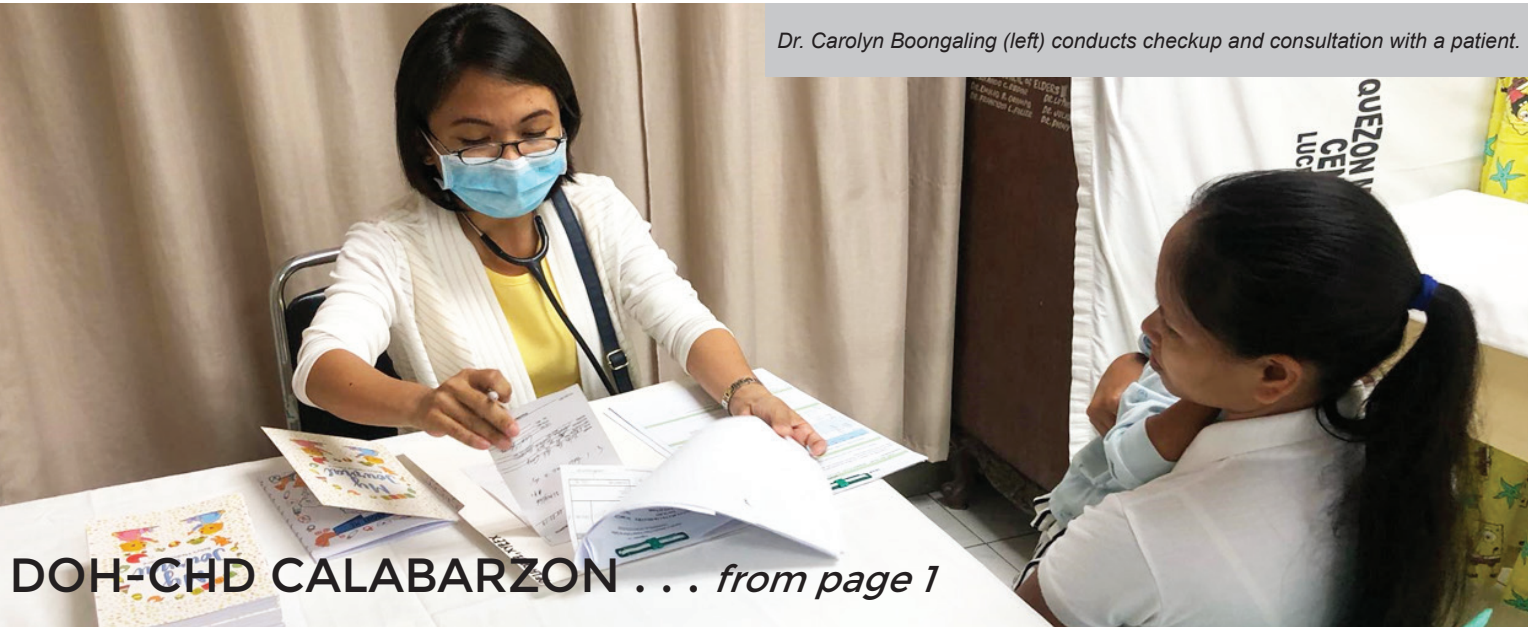
spearheaded by Julie Soria, provincial NBS coordinator.

Meanwhile, DOH-CHD Caraga held a similar ENBS training on August 28-29 at Shacene Royal Function Hall in Tandag City. The Adela Serra Ty Memorial Medical Center hosted the practicum on the second day. Also part of the program was the presentation of provincial and regional statistics on program coverage and quality indicators to evaluate the performance of each NSF.

Dr. Conchita G. Abarquez, NSC-M Unit Head, discussed the panel of disorders in Cagayan de Oro City, while Edbert Jasper M. Jover, NSC-M Genetic Counselor, discussed the same in Tandag City. **RAguilar**



DOH-CHD Caraga NBS Nurse Danile Montilla (first row, seated third from left) and NSC-M Genetic Counselor Edbert Jasper Kover with the participants from select health facilities in Caraga.



## DOH-CHD CALABARZON . . . from page 1

recalled, including the 21 patients that were tracked and home-visited during a span of three weeks conducted by Jose Antonio Yap and Lady Camille Delos Reyes, NBS coordinators of DOH-CHD CALABARZON; Dianne Marie Mejilla and Yancy Otioco, NBS coordinators of the Provincial Health Office of Quezon; and Jhonalyn Bantigue, Follow-up Nurse of the NBSCC. The remaining 12% of patients are still being recalled and/or are non-compliant to treatment and management, as some municipalities have limited access to health facilities and specialists and geographic terrain poses a challenge as well.

With the help of the different local government units (LGUs) in Quezon, all patients were found and categorized as indigents. Parents and guardians all signed their consent to participate in the said event. DOH-CHD CALABARZON sponsored the laboratory tests needed before the checkup, while the different

LGUs provided transportation to the venue.

The first PSPME follow-up clinic was set up and consultations conducted at the Quezon Medical Center (QMC), Lucena City, on August 22, 2019. Dr. Carolyn Boongaling and Dr. Jedeane Aragon discussed first the conditions of the patients to the parents and how they can take care of their child with these conditions. Free vitamins, dental kits, micronutrient supplements, and medicines were also given to the patients.

Dr. Dennis Rivere, head of the pediatric department of QMC, thanked the organizers for their support, especially in locating patients in their province. He also told the parents of the patients that they are taken care of by the NBS Program. In turn, the families promised to continue to take care of their children to the best of their ability. **JAYap**

## NSC-SL Organizes 2nd Thalassemia Forum

A total of 17 parents from the provinces of Batangas, Cavite, and Laguna attended the 2nd Thalassemia Forum for parents conducted by NSC-SL at Jollibee Calamba Crossing, Province of Laguna, on August 28, 2019.

Dr. Jocelyn R. Rosita, a pediatric hematologist-oncologist, served as the resource speaker. She discussed the nature of the disorders, the probabilities of inheritance, the tests necessary for each disorder, possible negative outcome if undetected and unmanaged, and the prognosis for the disorders.

The lecture was followed by the forum for parents in order to provide clarification on the general concerns they have of the disorder.

As the prevalence of these disorders under the hemoglobinopathies group increase, NSC-SL has drawn up plans to raise the forum to the provincial level to meet a wider audience. **FDimaculangan**

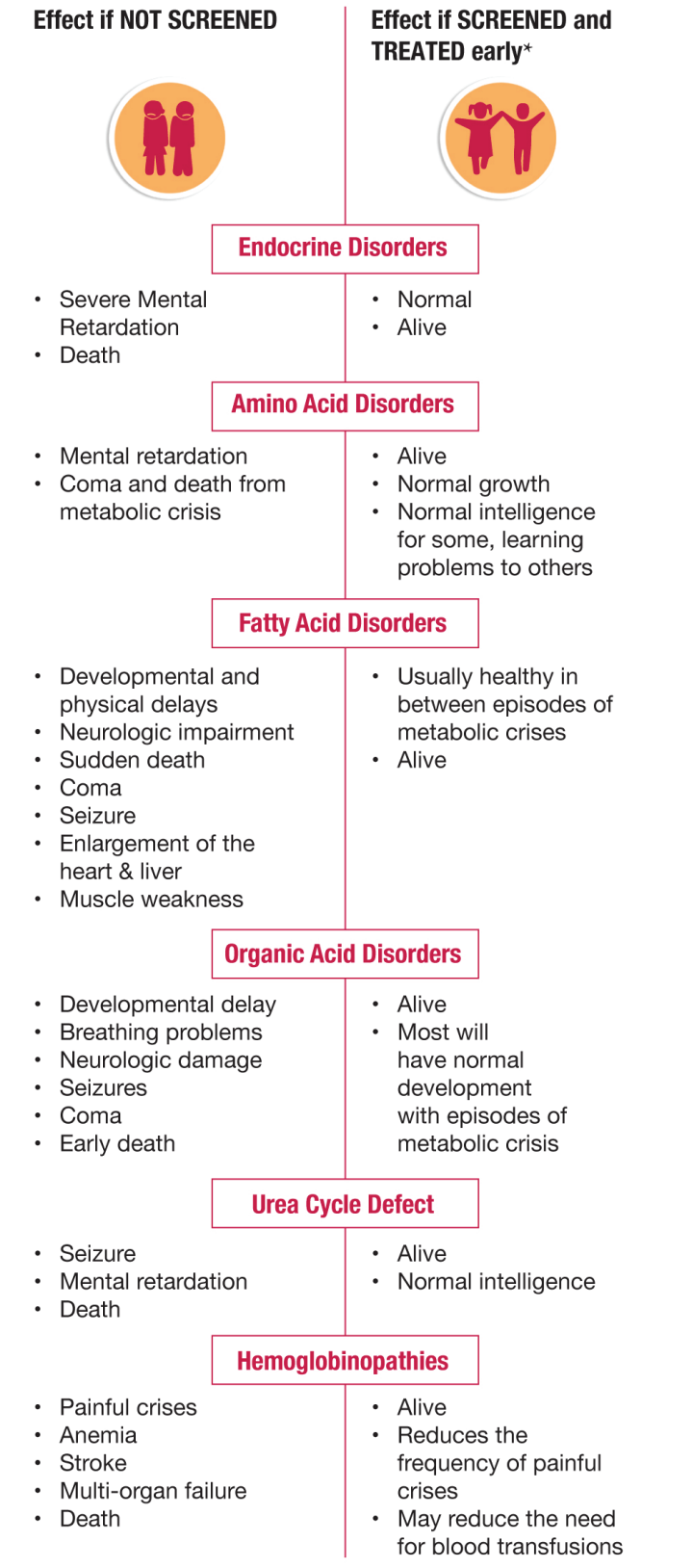


Attendees and organizers of the 2nd Thalassemia Forum in Calamba City.

# HELP US SAVE BABIES AFFECTED BY ANY OF THESE DISORDERS.

- Endocrine Disorders**
- Congenital Hypothyroidism
  - Congenital Adrenal Hyperplasia
- Amino Acid Disorders**
- Homocystinuria
  - Hypermethioninemia/Methionine Adenosine Transferase Deficiency
  - Maple Syrup Urine Disease
  - Phenylketonuria
  - Tyrosinemia Type I
  - Tyrosinemia Type II, III
- Fatty Acid Disorders**
- Carnitine Palmitoyltransferase I Deficiency
  - Carnitine Palmitoyltransferase II Deficiency
  - Carnitine Uptake Deficiency
  - Glutaric Acidemia Type II
  - Long Chain Hydroxyacyl-CoA Dehydrogenase Deficiency
  - Medium Chain-Acyl-CoA Dehydrogenase Deficiency
  - Very Long Chain-Acyl-CoA Dehydrogenase Deficiency
  - Tri-functional Protein Deficiency
- Organic Acid Disorders**
- 3-Methylcrotonyl CoA Carboxylase Deficiency
  - Beta Ketothiolase Deficiency
  - Glutaric Acidemia Type I
  - Isovaleric Acidemia
  - Methylmalonic Acidemia
  - Multiple Carboxylase Deficiency
  - Propionic Acidemia
- Urea Cycle Defect**
- Citrullinemia
  - Argininosuccinic Aciduria
- Hemoglobinopathies**
- Alpha Thalassemia
  - Beta Thalassemia
  - Hemoglobin C
  - Hemoglobin D
  - Hemoglobin E
  - Sickle Cell Disease
- Others**
- Galactosemia
  - Glucose-6-Phosphate Dehydrogenase Deficiency
  - Cystic Fibrosis
  - Biotinidase Deficiency

## WHY SCREEN YOUR BABIES?



\*Long-term follow-up and management of children with confirmed newborn screening conditions ensure that these children receive the full benefits of early identification through newborn screening.

## ANNOUNCEMENTS

The Philippines is hosting the 13th Asia-Pacific Conference on Human Genetics at the Makati Shangri-La Hotel on November 7-9, 2019, with the theme "Advancing Translational Medicine and Collaborations in Genomics." This conference will bring together an outstanding community of faculty, scientists, researchers, and students from the Asia-Pacific region and different parts of the world.

Simultaneous sessions are relevant to the National Newborn Screening Program such as Tracks on Revolutionizing Newborn Screening: New Developments and Dilemma; Advances in the Diagnosis and Therapy of Inherited Metabolic Diseases; and Hemoglobinopathies: Advances in Diagnosis and Novel Therapeutics. These will help the participants, especially those caring for patients diagnosed with any of the disorders in the expanded newborn screening program, be up-to-date.



## Newborn Screening Week: October 1 to 7

Learn more about the disorders included in the EXPANDED NEWBORN SCREENING PANEL!

www.newbornscreening.ph

#eNBSSaves



Every first week of October, the country is celebrating the National Newborn Screening Week by virtue of Presidential Proclamation No. 540. This week provides an excellent opportunity to celebrate the success of the program and to present the comprehensive newborn health program for the benefit of the newborns and their families.

In this year's celebration of the Newborn Screening Awareness Week, please join us again in letting the general public know about the benefits of newborn screening and the inclusion of expanded screening in PhilHealth's Newborn Care Package from October 1-7, 2019. Work with us in saving Filipino newborns from mental retardation and death!

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## IN THE KNOW HOMOCYSTINURIA

**What is Hcy?**  
 Homocystinuria (Hcy) is caused by cystathionine β-synthase deficiency, an inborn error of the transsulfation pathway that causes an increase in levels of homocysteine and methionine in the blood.

**Clinical Manifestation**  
 Patients affected with Homocystinuria may present with (1) ectopia lentis, which is found in 85% of patients; (2) skeletal abnormalities are prominent especially genu valgus and patients are often described to have a "marfanoid habitus"; (3) mental retardation is common but not invariable; and (4) thromboembolism.

**Pathophysiology**  
 Increased homocysteine levels is found to inhibit linking of collagen and elastic tissue, which predisposes zonule generation of the eye predisposing patients to myopia and lens dislocation. Skeletal abnormalities are thought to result from damage to fibrillin in patients with cyathionine β-synthase and due to a reduction in collagen crosslinking, while the mechanism that contributes to the atherogenic propensity of hyperhomocystinemia is related to endothelial dysfunction and injury, which leads to platelet aggregation and thrombus formation. Finally, chemical abnormalities and the repeated thromboembolic strokes may contribute to the mental retardation.

**Inheritance:** Autosomal recessive

**Confirmatory Testing**  
 Total homocysteine in plasma. Amino acids in plasma, methylmalonic acid in urine and enzyme study in fibroblasts may be used to confirm the diagnosis.

**Overview of Disease Management**  
 The aim of treatment is to reduce the plasma total homocysteine through the following approaches: (1) large doses of pyridoxine (50-100 mg/day) have been effective in reducing biochemical abnormalities in patients with cystathionine-β-synthase deficiency where about half respond partially; (2) folic acid (10 mg/day) may be given along with betaine (100 mg/kg/day) that lowers homocysteine levels by remethylation dietary modification by giving a low-methionine / high-cystine diet. Additional treatment may include Vitamin C (100 mg/day) and hydroxocobalamin (1 mg/day) starting at 5 years of age.

**Prognosis**  
 Early diagnosis and treatment can prevent thromboembolic events and reduce the complications brought about by increased levels of homocysteine.

**Preliminary / Initial Management During Metabolic Crisis**  
 Metabolic crises may be caused by illness, prolonged fasting, or stressful situations such as surgery and severe infection. The goal of treatment is to reverse the catabolic state, correct the acidosis, and prevent essential amino acid deficiency.

**What To Do**  
 If unwell and cannot tolerate oral intake:  
 · Nothing per ore  
 · Ensure patient's airway is secure  
 · Insert IV access. Collect samples for plasma amino acids.  
 · May request for investigations (i.e., CBC etc.) as needed.  
 · May give fluid boluses if patient requires.  
 · Start D12.5% 0.3NaCl at full maintenance. Assess patient clinically; if there is need to increase fluid, may do so up to 1.2 or 1.5x the maintenance.  
 · Start betaine, folic acid, and vitamin B6.  
 · Monitor input and output strictly (q6 hours).

If unwell and can tolerate oral intake:  
 · Insert oro- or nasogastric tube and start continuous feeding with Hcy formula or protein free formula at maintenance rate  
 · Insert IV access. Collect samples for plasma amino acids.  
 · May request for investigations (i.e., CBC etc.) as needed.  
 · Start D12.5% 0.3NaCl at 5-10 cc/hr  
 · Start betaine, folic acid, and vitamin B6.  
 · Monitor input and output strictly (q6 hours)

\*Children should not be protein-restricted for longer than necessary (24-28 hours). Inform metabolic doctor on call for further guidance regarding ongoing management.

## IPHO . . . from page 3

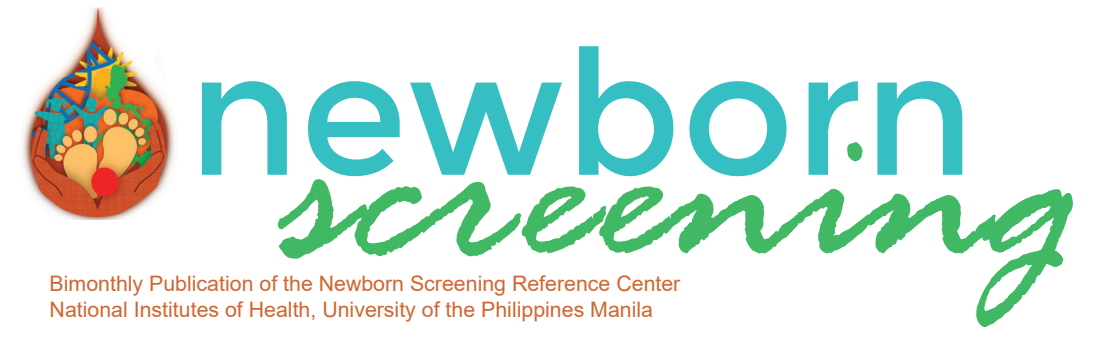
One of the highlights of the activity was the gallery exhibits where different rural health units from different municipalities presented their accomplishments in their health programs, including newborn screening.

Amerah Matuan, who represented NSC-Mindanao (NSC-M), presented the updates on the Expanded Newborn Screening (ENBS) Program. Matuan also provided guidelines for newborn screening facilities (NSFs) on how to implement ENBS in their respective stations and provided materials to assist them in their advocacies.

The event also served as a venue for all the NSF coordinators to raise concerns and issues encountered in their workplace such as logistics, PhilHealth claims, actual implementation, and cost for patients' confirmatory testing. *AMatuan*

Thank you for reading our newsletters!  
 We would love to hear your feedback. Please let us know how we can improve our bimonthly newsletters by answering our feedback form at [www.newbornscreening.ph](http://www.newbornscreening.ph).  
 If you would like to write an article, please contact us at [info@newbornscreening.ph](mailto:info@newbornscreening.ph).  
 We will do our best to keep you informed about current and relevant newborn screening issues.

**Your Feedback Is Important To Us**



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## DOH-CHD CALABARZON Sets Up Free Clinic, Consultations

The Newborn Screening Continuity Clinic (NBSCC) Region CALABARZON, hosted by General Emilio Aguinaldo Memorial Hospital, has in its record 52 patients with Congenital Hypothyroidism (CH) and 9 patients with Congenital Adrenal Hyperplasia (CAH) detected through the newborn screening program for the entire province of Quezon. This does not represent the entire number, as more cases are still being endorsed by the Newborn Screening Centers (NSCs).

For this reason, the Department of Health-Center for Health Development (DOH-CHD) CALABARZON and NBSCC Region 4A decided to conduct its first Philippine Society of Pediatric Metabolism and Endocrinology (PSPME) follow-up consultation in this area. This free consultation service provided by pediatric endocrinologists aimed at assessing patients' health, growth, and development in relation to the patients' endocrine disorders. This activity is part of enhancing long-term management and patient care in the region.

Of the total 61 patients with CH and CAH, 46 (88%) were  
*Continued on page 5 . . .*

Number of Babies Screened as of August 2019 : **12,238,604**

## What's Inside

2. NSC-NL Passes Reaccreditation
3. PHO Benguet Conducts Common Metabolic Disorders Forum • IPHO-Maguindanao Hold 2019 Semi-Annual PIR
4. Mindanao CHDs, NSC-M Hold NBS Trainings
5. NSC-SL Organizes 2nd Thalassemia Forum
7. Announcements
8. In The Know Homocystinuria