NSC NIH
NSC-NIH: For Regions IVB, V & NCR
National Institutes of Health, Bldg. H
UP Ayala Land Technohub Complex
Diliman, Quezon City
Telephone: (02) 376-0962, 376-0967
Fax: (02) 921-6395
Email: nsc-nih@up.edu.ph

DOH-NCRO Holds G6PD Deficiency Forum

Participants and organizers share a group photo together after the forum in Hotel H20, Manila.

In collaboration with NSC-NIH, DOH-NCRO conducted a G6PD Deficiency Forum at Hotel H2O in Manila on August 17, 2018.

The forum is a regular activity to increase awareness among the public about Glucose-6-Phosphate Dehydrogenase (G6PD) Deficiency, a prevalent condition in the country affecting 1 in every 58 newborns. Individuals with this deficiency may suffer from hemolytic anemia resulting from exposure to oxidative substances found in drugs, foods, and chemicals.

Children with G6PD Deficiency and their parents and guardians attended the activity together with other health partners and stakeholders. Dr. Maria Teresa B. Rivera, Family Health Cluster Head of DOH-NCRO, welcomed the participants.

Two distinguished speakers were invited to talk about the disorder. Dr. Maria Beatriz Gepte, NBS Chair of the G6PD Deficiency Expert Panel and a pediatric hematologist and oncologist from the Philippine Children's Medical Center, discussed the clinical aspects of G6PD Deficiency, while Dr. Catherine Lynn Silao, a geneticist and Molecular Genetics Laboratory Unit Head at the Institute of Human Genetics, University of the Philippines Manila, discussed the genetic aspects of the disorder.

An open forum was held after each discussion to allow the parents to pose their questions pertaining to their children's condition. *CAlincastre*

DOH-NCRO . . . *from cover*

program implementers and other families present.

Children, on the other hand, were treated to a magic show and face painting activities. Some members of the Volunteer Youth Leaders for Health (VYLH) also led several games. *CAlincastre*

Top: A magician performs some tricks for the kids during the magic show. Bottom: VYLH volunteers facilitate fun activities and games for children and parents.

Mindanao NSC
For Mindanao
Southern Philippines Medical Center
J. P. Laurel Avenue, Davao City
Telephone: (082) 226-4595, 224-0337
Telefax: (082) 227-4152
Email: nbsprogram@nscmindanao.ph

Participants of the ENBS training and orientation at Paraiso Verde Resort, Koronadal City.

DOH-RO 12 Conducts 2nd Leg of ENBS Training

To ensure better implementation and increase newborn screening coverage, DOH-RO 12 conducted the second leg of its ENBS Training for the year at Paraiso Verde Resort, Koronadal City, South Cotabato, on July 10 and 11, 2018.

The ENBS panel of disorders, administrative mechanics of NSC-Mindanao (NSC-M), and types of unsatisfactory samples were discussed by Dr. Conchita Abarquez, NSC-M Unit Head, and Perly Bermudez, Project Development Officer. Mary Agnes Panton, NBS Program Manager of DOH-RO 12, highlighted the significance and legal aspects of the NBS.

The dos and don'ts of NBS sample collection were explained by Rohainnah Mua, Regional Nurse Coordinator, while long-term patient care and the role of the NBS Continuity Clinics in the program was expounded by Juledene Mendoza, follow-up nurse.

Continued on page 4 . . .

IMAP Region 11 Taps NSC-M for ENBS Updates

"The benefits of expanded newborn screening outweigh the cost," explained Dr. Conchita Abarquez, NSC-M Unit Head, during the Clinical Case Conference for Midwives organized by the Integrated Midwives of the Philippines (IMAP) in Tagum City on August 23-24, 2018.

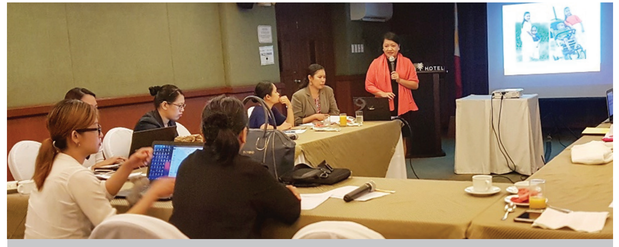
Dr. Abarquez was invited by the IMAP Region 11 Chapter to provide technical information and updates on ENBS. With its implementation in Mindanao, health professionals, especially midwives, involved in newborn care have an urgent need for knowledge on this type of screening. The midwives in Davao Regions were no exception. Further, some of the midwives were still coming to terms on the cost of ENBS.

After the presentation, midwives were able to ask questions regarding ENBS implementation in their respective facilities, financial support for patients, and protocols on repeating

Continued on page 8 . . .

Central Luzon NSC
For Regions III & CAR
Angeles University Foundation Medical Center
MacArthur Highway, Barangay Salapungan,
Angeles City
Telephone: (045) 624-6571
Email: nscaufmc@gmail.com

NSC-CL Conducts Annual Consultative Meeting



The resource speaker engages the participants of the consultative meeting in a discussion at Oasis Hotel, Angeles City.

To continually improve program implementation and coverage, NSC-Central Luzon (NSC-CL) fortified its partnership with the NBS Teams of DOH-ROs in Central Luzon and the Cordillera Administrative Region through an annual consultative meeting held at the Oasis Hotel, Angeles City, on July 31, 2018.

Representatives from the Newborn Screening Reference Center (NSRC) were also present to give inputs to the group's plans for the remainder of the year.

Consultative meetings, which are an essential part of the NBS Program, gather together representatives from the NSCs, DOH-ROs, and NSRC to discuss accomplishments, plans of activities, and other relevant issues. It also serves as a venue to share insights, improve strategies, solve problems, and widen ventures toward the achievement of a common goal—to provide quality newborn screening services to all Filipino babies. *NDelaCruz*

DOH-RO 12 . . . *from page 3*

On the second day of the training, the participants underwent a practicum on heel pricking at South Cotabato Provincial Hospital in Koronadal City and were assisted by Merly Feria and other hospital NBS team members.

The two-day event was attended by 27 health professionals from government and private health facilities in South Cotabato, Sultan Kudarat, Cotabato City, General Santos City, and North Cotabato. *RMua, PBermudez*

A participant tests her skills in heel prick practicum.

NSC Southern Luzon
For Region IVA
Daniel O. Mercado Medical Center
3rd Flr. DMMC-HIS Bldg., 143 Narra St.,
Mountview Subd., Tanauan City
Telephone: (043) 702-7715, 702-7719
Email: nsc.southernluzon@gmail.com

Top: Organizers of the G6PD Forum join the babies with G6PD Deficiency and their parents and guardians in Cavite. Bottom: Babies and their parents and guardians from Batangas and Laguna provinces pose for a group photo with the forum organizers.

NSC-SL Holds Back-to-Back G6PD Fora

To educate the parents of newborns with G6PD Deficiency as well as organize a support group in Southern Luzon, NSC–Southern Luzon (NSC-SL) conducted a back-to-back G6PD Deficiency Parents' Forum for Cavite and Laguna-Batangas on August 14 and 16, 2018, respectively.

In Cavite, Dr. Michael C. Cuarteron, a pediatric hematologist and oncologist, served as guest speaker and gave a lecture titled "7 Most Commonly Asked Questions About G6PD Deficiency." Dr. Cuarteron discussed the disorder in detail to the 57 participants, including the genetic aspects, manifestations, and some recommendations for the patients with G6PD Deficiency. Meanwhile, in Batangas and Laguna, Dr. Jocelyn R. Rosita, also a pediatric hematologist and oncologist, spoke to 64 parents in attendance.

At the end of the lecture, parents were given the chance to raise their questions. NSC-SL also took the opportunity to help indigent patients who were initially screened positive for G6PD Deficiency by providing free confirmatory tests, which were availed of by 46 babies from the three provinces. *FDimaculangan*

Get your newborn screened!

EXPANDED NEWBORN SCREENING IS NOW AVAILABLE

Visit www.newbornscreening.ph or ask your health attendant about expanded newborn screening.

PhilHealth Your Partner in Health

IN THE KNOW Very Long-Chain Acyl-CoA Dehydrogenase Deficiency

7

Very Long-Chain Acyl-CoA Dehydrogenase (VLCAD) catalyzes the dehydrogenation of C22-C12 straight chain fatty acids. And because the long chain fatty acids constitute a major proportion of the fatty acids, VLCAD Deficiency is generally a more severe condition than MCAD or SCAD deficiency, and multiple tissues are affected.

Clinical Manifestation. The clinical presentation of symptomatic VLCAD Deficiency is heterogenous with phenotypes of different severities. There are three forms described: (1) severe childhood form with neonatal onset and cardiomyopathy; (2) milder childhood form with delayed onset of symptoms often triggered by metabolic stress and presents as hypoketotic hypoglycemia; and (3) adult form which presents with isolated skeletal muscle involvement with recurrent episode of muscle pain, rhabdomyolysis, and myoglobinuria.

Pathophysiology. VLCAD catalyzes the dehydrogenation of acyl CoA esters of 14-20 carbon length in the first step of mitochondrial fatty acid oxidation. VLCAD Deficiency results in lack of production of energy from β-oxidation of long-chain fatty acids, because heart and muscle tissue depend heavily on energy from long chain fatty acid oxidation; a VLCAD Deficiency severely affect these tissues.

Inheritance: Autosomal recessive

Confirmatory Testing. The enzyme defect can be detected through culture skin fibroblasts. The gene for VLCAD has been cloned and sequenced successfully and plays a role in diagnosis of this disorder.

Overview of Disease Management. Treatment of this disorder includes avoidance of fasting by frequent feeding, overnight continuous feeding, reduction in the amount of long chain fat in diet while supplying essential fatty acids in the form of canola oil, walnut oil, or safflower oil, and supplementation with medium chain triglycerides.

For the adult muscular form, it is advised to have a high carbohydrate intake prior to exercise to prevent lipolysis and to restrict physical activity to levels that are not likely to precipitate an attack of rhabdomyolysis.

Prognosis. Fifty percent of patients die within 2 months of initial symptomatology. However, timely and correct diagnosis leads to dramatic recovery so that early detection could prevent the onset of arrhythmias, heart failure, metabolic insufficiency, and death.

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:
a. Nothing per orem
b. Ensure patient's airway is secure
c. Insert IV access. Monitor glucose levels. Collect samples for urine ketones and serum creatine kinase (CK). May request for investigations (i.e., CBC, liver transaminases, blood gas, etc.) as needed.
d. May give fluid boluses if patient requires.
e. Start D10% 0.3NaCl at full maintenance. Assess patient clinically; if there is need to increase fluid, may do so up to 1.2x or 1.5x the maintenance.
f. Monitor input and output strictly (q6 hours). Check color of urine and may request for urinalysis to check for urine myoglobin.

If unwell and can tolerate oral intake:
a. Encourage regular feeding.
b. Insert IV access. Monitor glucose levels. Collect samples for urine ketones and serum creatine kinase (CK). May request for investigations (i.e., CBC, liver transaminases,

Continued on page 8 . . .

EDITORIAL STAFF

Executive Editor: Ma. Elouisa L. Reyes
Managing Editor: Vina G. Mendoza
Associate Editor: Emerson Kim J. Lineses
Layout Artist: Emerson Kim J. Lineses
Contributors: Claire Alincastre, Perly Bermudez, Nikki Dela Cruz, Frex Dimaculangan, Sheila Mae Guilaran, Rohainnah Mua
Circulation: Marjorie Dandan, Jesusa Jalad, Juvelyn Reyes, Ma. Agnes Algarme, Felix Alipasa
Adviser: Riza Concordia N. Suarez

Comments and questions may be sent to:

Newborn Screening Reference Center
National Institutes of Health
University of the Philippines Manila
Unit 304 New Gold Bond Building
1579 F. T. Benitez, Ermita, Manila

Or email us at: info@newbornscreening.ph
Website: www.newbornscreening.ph
Follow us on Twitter: @newbornscreenph

8

Left: Dr. Abarquez delivers her presentation on the ENBS program in the Philippines to the members of IMAP Chapter 11. Right: Participants read and examine educational materials on ENBS, which were given to them for free.

VLCAD Deficiency . . . *from page 7*

blood gas, etc.) as needed.
c. Start D10% 0.3NaCl at 5-10 cc/hr.
d. Monitor input and output strictly (q6 hours). Check color of urine and may request for urinalysis to check for urine myoglobin.

*Inform metabolic doctor on call for further guidance regarding ongoing management.

Source: Fact Sheets for Doctors, May 2016, accessed at www.newbornscreening.ph on August 4, 2018.

IMAP Region 11 . . . *from page 3*

newborn screening for preterm babies, the ideal period for screening newborns, sample collection, and card replacements.

Meanwhile, IMAP members in the Davao regions expressed support by increasing the availability of ENBS in lying-in clinics and birthing homes managed by IMAP. *SMGuilaran*

Your Feedback Is Important to Us!

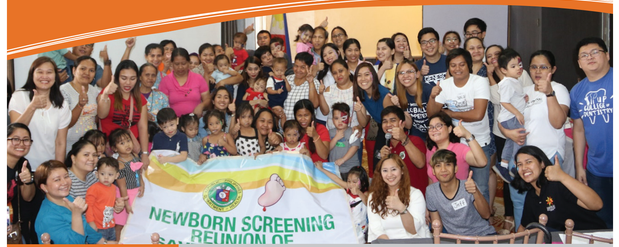
Thank you for reading our newsletters!
We would love to hear your feedback. Please let us know how we can improve our bi-monthly newsletters by answering our feedback form at www.newbornscreening.ph. If you would like to write an article, please contact us at info@newbornscreening.ph. We will do our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

Newborn Screening Reference Center
National Institutes of Health
University of the Philippines Manila
Unit 304 New Gold Bond Building
1579 F. T. Benitez, Ermita, Manila

VOLUME 22
ISSUE 4
JULY-AUGUST 2018

newborn screening

Bimonthly Publication of the Newborn Screening Reference Center
National Institutes of Health, University of the Philippines Manila



Top: Participants and organizers of the reunion. Bottom. A parent of children screened positive for MSUD shares her gratitude for the newborn screening program.

What's Inside

2 DOH-NCRO Holds G6PD Deficiency Forum
3 DOH-RO 12 Conducts 2nd Leg of ENBS Training
IMAP Region 11 Taps NSC-M for ENBS Updates
4 NSC-CL Conducts Annual Consultative Meeting
5 NSC-SL Holds Back-to-Back G6PD Fora
7 Very Long-Chain Acyl-CoA Dehydrogenase (VLCAD) Deficiency

DOH-NCRO Conducts Reunion of Saved Babies

"Nagpapasalamat ako sa newborn screening dahil nakita agad ang kalagayan ng mga bata maaga pa lang," shared a parent of patients with Maple Syrup Urine Disease (MSUD) during the Reunion of Saved Babies conducted by the Department of Health–National Capital Regional Office (DOH-NCRO) at Greenhills Elan Hotel Modern, San Juan City, on July 27, 2018.

In collaboration with the Newborn Screening Center–National Institutes of Health (NSC-NIH), the event gathered together children aged 3 to 5 years accompanied by their parents and guardians along with program partners and stakeholders.

Dr. Anna Lea G. Elizaga, Unit Head of NSC-NIH, gave a short talk on Expanded Newborn Screening (ENBS) to help the parents understand more about the additional disorders included in the panel and the benefits of early screening. Meanwhile, Mikko Myron De Guzman, Follow-Up Nurse at the Philippine General Hospital Continuity Clinic, discussed the importance of long-term follow-up care.

The video of the 20th year of Newborn Screening Program was also presented, which featured testimonials from families and children saved through newborn screening. Parents and guardians were also able to express their gratitude to the

Continued on page 2 . . .

Number of Babies Screened as of August 2018 : 11,648,378