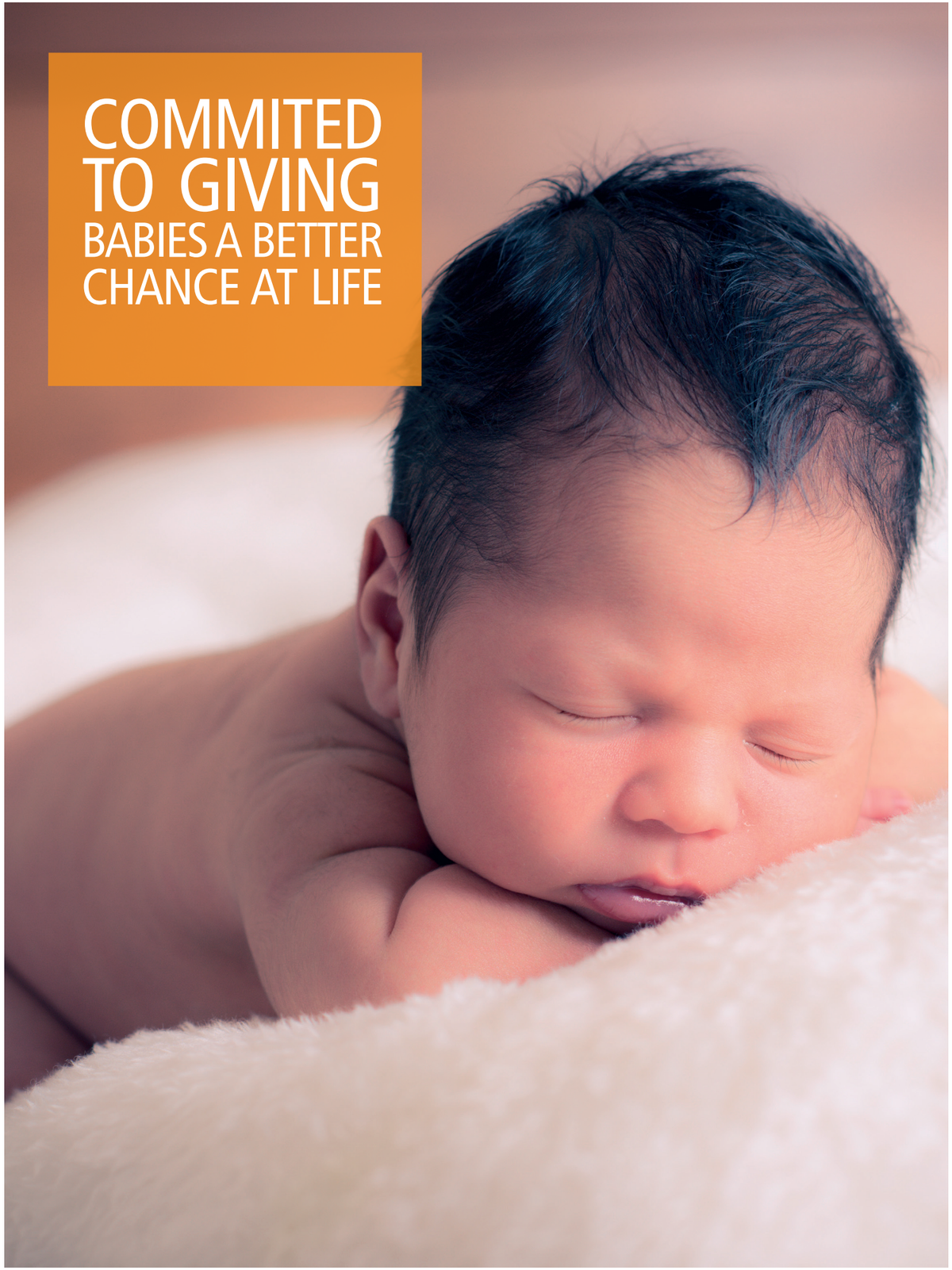
NeoBase™ 2 Non-derivatized MSMS kit

The next generation NeoBase™ 2 Non-derivatized MSMS kit gives you the ability to screen for all the common amino acid, organic acid and fatty acid oxidation disorders as well as purine and peroxisomal metabolic disorders.

PerkinElmer®
For the Better



NEOBASE™ 2 KIT, THE NEXT GENERATION IN MSMS BASED NEWBORN SCREENING

PerkinElmer, a pioneer and the global leader in mass spectrometry based newborn screening since 2002, currently offers the NeoBase™ Non-derivatized MSMS Kit, the gold standard in mass spectrometry based testing for amino acid, organic acid, and fatty acid oxidation disorders, used to screen for millions of babies annually.

While the NeoBase™ kit has serviced the industry well, we have collected feedback from customers over the last decade to determine how we can make it easier for labs to test for more disorders, with less resources, while reducing their false positive rate. Based upon requests to be able to screen for more disorders in less time from a single punch in a simple three step workflow, we are proud to be developing the NeoBase™ 2 kit, the next generation non-derivatized MSMS kit, the efficient and cost-effective way to expand newborn screening without the need for additional equipment and resources.

WHY CHOOSE NEOBASE™ 2 FOR EXPANDED SCREENING?

- ☑ **Test for more than 50 analytes, including markers for screening of X-ALD and ADA SCID, from a single DBS punch with a single extraction and incubation step**
- ☑ **A reformulated Flow Solvent that improves ionization and makes it easy to switch between the NeoLSD™ kit and NeoBase™ 2 kit**
- ☑ **DBS quality controls with analyte concentrations adjusted at clinically significant levels**
- ☑ **All internal standards are in a single vial making reconstitution easy. Extraction solution is stable for up to two weeks, eliminating the need for daily preparation.**
- ☑ **Shorter extraction time, shorter incubation time and shorter run time**
- ☑ **New specific markers for Urea Cycle Disorders ASA-LD, OTCD, CPS-I and NAGS as well as for ADA SCID providing more conclusive screening results**

FASTER AND SIMPLER ASSAY WORKFLOW

KIT INCLUDES ALL COMPONENTS YOU NEED FOR EFFECTIVE NEWBORN SCREENING

20 U-bottomed microplates
ANSI-standard U-bottomed microplates

20 sheets of adhesive microplate covers
Easy-to-use adhesive covers for all assay steps

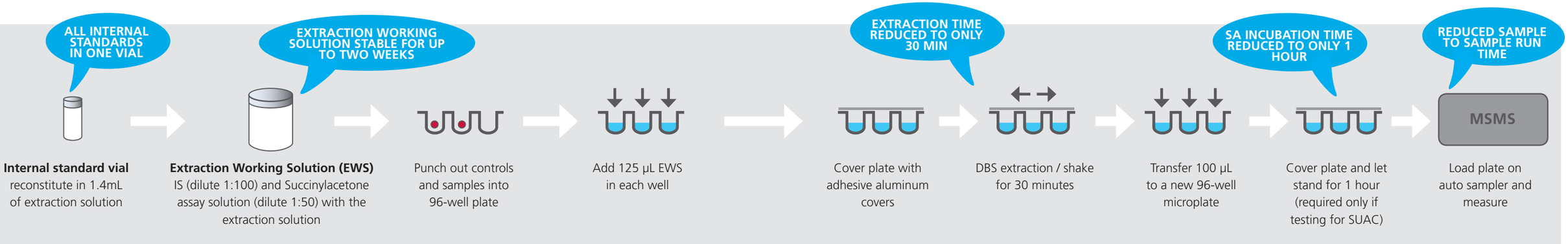
NeoBase™ 2 internal standards
All 30 internal standards provided in a single vial uptime

Controls
Kit controls, Low and High. Three filter paper cassettes containing 3 spots of each level per cassette..

Lot specific quality control certificate
The exact concentrations of isotope labeled internal standards measured by the manufacturer are given on the lot-specific quality control certificate included in the kit

NeoBase™ 2 Non-derivatized Assay Solution
The package includes a bottle of each of extraction solution and flow solvent. One package is enough for two NeoBase™ 2 Non-derivatized MSMS kits.

NeoBase™ 2 Succinylacetone Assay Solution
optional reagent for the detection of succinylacetone when screening for tyrosinemia type I is desired



Internal standard vial
reconstitute in 1.4mL of extraction solution

Extraction Working Solution (EWS)
IS (dilute 1:100) and Succinylacetone assay solution (dilute 1:50) with the extraction solution

Punch out controls and samples into 96-well plate

Add 125 µL EWS in each well

Cover plate with adhesive aluminum covers

DBS extraction / shake for 30 minutes

Transfer 100 µL to a new 96-well microplate

Cover plate and let stand for 1 hour (required only if testing for SUAC)

Load plate on auto sampler and measure

MORE ANALYTES, IMPROVED PERFORMANCE, BETTER PRODUCTIVITY

Table 1. Analytes measured by the NeoBase™ 2 Non-derivatized MSMS Kit and their corresponding internal standards and controls.

AMINO ACIDS		
ANALYTE	INTERNAL STANDARDS	CONTROLS
Ala	$^{2}H_{3}$-Ala	Ala
Arg	$^{2}H_{4}$,^{13}C-Arg	Arg
Asa*	$^{2}H_{4}$,^{13}C-Arg	Asa
Cit	$^{2}H_{2}$-Cit	Cit
Gln	$^{13}C_{5}$-Gln	Gln
Glu	$^{13}C_{5}$-Gln	Gln
Gly	^{15}N,2-^{13}C-Gly	Gly
Leu/Ile/Pro-OH	$^{2}H_{3}$-Leu	Leu
Met	$^{2}H_{3}$-Met	Met
Orn	$^{2}H_{6}$-Orn	Orn
Phe	$^{13}C_{6}$-Phe	Phe
Pro	$^{13}C_{5}$-Pro	Pro
Tyr	$^{13}C_{6}$-Tyr	Tyr
Val	^{15}N-$^{13}C^{5}$-Val	Val

*Asa is measured as a total concentration of Asa and its anhydrides.

KETONES		
ANALYTE	INTERNAL STANDARDS	CONTROLS
SA	$^{13}C_{5}$-MPP**	SA

** Stable-isotope labeled succinylacetone (SA) derivative 3-(5-methyl-1H-pyrazol-3-yl) propanoic acid (MPP).

NUCLEOSIDES		
ANALYTE	INTERNAL STANDARDS	CONTROLS
ADO	$^{13}C_{5}$-ADO	ADO
dADO	$^{13}C_{5}$-dADO	ADO

PHOSPHOLIPIDS		
ANALYTE	INTERNAL STANDARDS	CONTROLS
C20:0 LPC*	2H4-C26:0 LPC	C26:0 LPC
C22:0 LPC*	2H4-C26:0 LPC	C26:0 LPC
C24:0 LPC	2H4-C26:0 LPC	C26:0 LPC
C26:0 LPC	2H4-C26:0 LPC	C26:0 LPC

* Not included in the US specific kit version 3044-001U.

CARNITINES		
ANALYTE	INTERNAL STANDARDS	CONTROLS
C0	2H9-C0	C0
C2	2H3-C2	C2
C3	2H3-C3	C3
C3DC/C4OH	2H3-C4	C4
C4	2H3-C4	C4
C4DC/C5OH	2H9-C5	C5
C5	2H9-C5	C5
C5:1	2H9-C5	C5
C5DC/C6OH	2H6-C5DC	C5DC
C6	2H3-C6	C6
C6DC	2H6-C5DC	C5DC
C8	2H3-C8	C8
C8:1	2H3-C8	C8
C10	2H3-C10	C10
C10:1	2H3-C10	C10
C10:2	2H3-C10	C10
C12	2H3-C12	C12
C12:1	2H3-C12	C12
C14	2H3-C14	C14
C14:1	2H3-C14	C14
C14:2	2H3-C14	C14
C14OH	2H3-C14	C14
C16	2H3-C16	C16
C16:1	2H3-C16	C16
C16OH	2H3-C16	C16
C16:1OH	2H3-C16	C16
C18	2H3-C18	C18
C18:1	2H3-C18	C18
C18:2	2H3-C18	C18
C18OH	2H3-C18	C18
C18:1OH	2H3-C18	C18
C18:2OH	2H3-C18	C18
C20*	2H3-C26	C26
C22*	2H3-C26	C26
C24*	2H3-C26	C26
C26*	2H3-C26	C26

IMPROVED FOR EVEN BETTER EXPANDED SCREENING

Main new features compared to the NeoBase™ Non-derivatized kit are:

- **New analytes: amino acids (ASA,Glu, Gln), acylcarnitines (C18:2OH, C20- C26*), lysophospholipids (LPC20:0*, LPC22:0*, LPC24:0, LPC26:0), nucleosides (Ado, dAdo)**
- **New single all-in-1 Internal Standards vial: Easier preparation and improved extraction working solution stability (2 weeks instead of 1 day)**
- **New kit controls with 2 optimized levels (Low, High)**
- **New single ANSI-standard plate and easy-to-use adhesive plate cover for all assay steps**
- **Non-derivatized Assay Solutions: New high quality borosilicate glass bottles, new Flow Solvent with lower MSMS background**
- **Succinylacetone Assay Solution: Faster succinylacetone (SA) extraction step with 60 min shorter waiting time**

OPTIONAL: MSMS WORKSTATION SOFTWARE

Complete new software for managing,reviewing and reporting results

KEY FEATURES:

- **Import data from instrument software**
  - Concentrations, IS intensties and TIC raw data
- **Visualize results**
  - Plate map with colored wells
  - Flagging of wells violating cutoffs or other QC criteria
  - View TIC & spectra
  - View results by disorder
  - Split grid view for easy review of results
- **SQL server database to store and consolidate data from one or more MSMS instruments**
- **Perform QC trend analysis and reporting**
- **Sample and assay audit tracking**
- **Automatic export to R4S for analysis**
- **Export results to LIMS**

COMPLETE MASS SPECTROMETRY SOLUTIONS FOR NEWBORN SCREENING. A COMMUNITY OF SUPPORT

From dried blood spot cards, punchers, instruments, reagents to informatics, PerkinElmer's mass spectrometry solutions empower newborn screening laboratories across the globe to meet their demands.

By joining the PerkinElmer newborn screening community, you become part of a movement that includes the majority of the newborn screening laboratories and leaders worldwide. In addition to all necessary equipment, reagents and informatics, we also provide on-site installation and training, ongoing support, phone consults and various training courses. We are here to help you improve your newborn screening program.

CONSIDERING EXPANDING YOUR SCREENING PANEL?

Why screen for X-linked adrenoleukodystrophy?

X-linked adrenoleukodystrophy (X-ALD) occurs when certain fats (very long chain fatty acids, or VLCFAs) cannot be broken down in the body. This disease largely affects the nervous system and the adrenal glands. When an individual has X-ALD, the buildup of VLCFAs may disrupt the fatty covering (myelin) of the nerve cells in the brain and spinal cord causing the myelin to breakdown. Without myelin, the nervous system cannot function properly causing for example difficulties swallowing or weakness in the legs. However, these symptoms vary depending on the type and age of onset and other factors which are not well understood. In addition, the build of VLCFAs damages the outer layer of the adrenal glands (adrenal cortex).

Prevalence

X-ALD affects 1:17,000 to 1:50,000 males and females (ref. Theda et al. Molecular Genetics and Metabolism, 111 (2014) worldwide. X-ALD affects males more severely and is more common in males. However, 20-40% of women who are carriers have symptoms in adulthood.

Why screen for ADA SCID?

Adenosine deaminase (ADA) deficiency is a systemic purine metabolic disorder caused by a defect in the enzyme adenosine deaminase. ADA deficiency can lead to severe combined immunodeficiency disease (SCID) which is usually fatal in infancy because of severe recurrent infections. Newborns with SCID may seem healthy at first because their mother's immune system protects them from infections for the first few weeks of life. However, without necessary treatment, common infections and vaccines can be life threatening. When diagnosis is made, permanent damage caused by infections or by metabolites is often present. Gene therapy, bone marrow transplantation, or enzyme therapy might be effective and life saving if performed early.

Prevalence

SCID affects 1 in 60,000 individuals worldwide (ref. van der Spek at al. J Clin Immunol, 35:4 (2015). About 15% of all SCID cases are due to adenosine deaminase deficiency.

Learn more on X-ALD and ADA SCID visit: www.newbornscreening.perkinelmer.com

NeoBase™ 2 MSMS kit may not be available in all countries, such as Canada. Please check out availability with your local PerkinElmer representative

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For a complete listing of our global offices, visit www.perkinelmer.com/ContactUs

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