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The Uneasy Truth Regarding the Ingredients in All Natural Blended Formulas

To the Editors: Within the past decade, the desire to feed ''real food'' to children with enteral tubes has led to a movement away from standard formulas and toward more blended tube feeds. Families perceive many nutritional benefits and like the ability to conform to specific dietary preferences (1). As more families choose to create their own pureed feeds, the commercial market for such feeds has exploded. Formula companies have developed shelf-stable, prepackaged, nutritionally complete blended formulas.

One family using such a commercially available ''natural'' formula had the unfortunate experience of finding their 4-year-old's gastrostomy tube clogged by a Cricket! (Fig. 1). Halfway through the child's feed the pump started alarming. This prompted them to look for a downstream occlusion and led them to discover that a full cricket had become lodged in the tubing of the gastrostomy tube. Nursing staff was forced to cut the extension tubing to extract the critter.

Despite regulations for the production of such blended feeds, and the fact that many of them are produced in the same facilities as standard formula, we must remain diligent in ensuring the quality. In the past, formulas have been recalled for similar incidences as described above (2). But with all natural, fresh fruits and vegetables, which are part of the agricultural process, is the risk of such quality control errors higher than before? If we are not careful, will the addition of more ''natural'' sources of protein, including crickets, become more common?

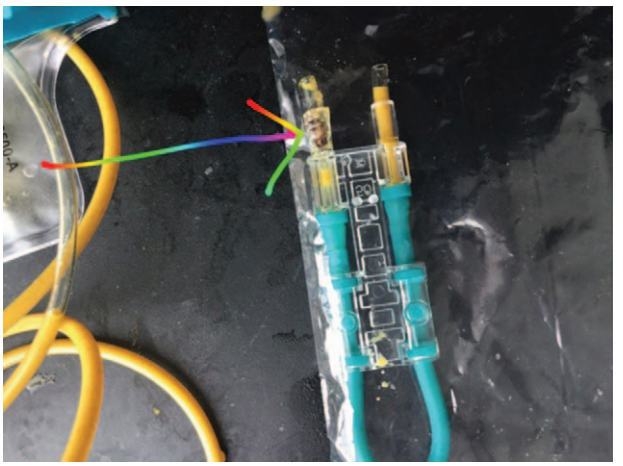

FIGURE 1. Full cricket found in feeding tube.

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Glycerol Phenylbutyrate Therapy in Progressive Familial Intrahepatic Cholestasis Type 2

To the Editor: In vitro studies have shown that sodium phenylbutyrate (PBA) can partially correct the misstrafficking of some endoplasmic reticulum–retained bile salt export pump missense mutants (1,2). In vivo studies in progressive familial intrahepatic cholestasis type 2 (PFIC2) patients carrying such mutants and treated orally with PBA (Ammonaps) have provided encouraging results with evidence for cholestasis improvement (1,2). Because of poor palatability and presentation, PBA administration and long-term adherence may be difficult (3). In addition, caution was made regarding possible hepatotoxicity due to interactions between PBA and rifampicin (4).

After in vitro studies showing that the p.G982R endoplasmic reticulum–retained bile salt export pump missense mutant was partially retargeted to the canalicular membrane with PBA, a PFIC2 girl carrying heterozygous compound *ABCB11* mutations (p.A257 V; p.G982R) was treated with PBA from 10 to 14.5 years (PBA: $275\,mg \cdot kg^{-1} \cdot day^{-1}$; 12 g/day, 24 tablets/day) (1). PBA treatment allowed disappearance of pruritus and normalization of serum bile acid (sBA) concentration (patient 2 (1)). Rifampicin was progressively tapered down then stopped and no evidence for PBA hepatotoxicity was found (4). Between 15 and 17 years she took PBA very intermittently due to difficulties with its administration, the consequence being periods with pruritus and cholestasis recurrence (sBA: 4–252 μmol/L; $N < 10$). Then, the patient decided to totally stop PBA. She experienced unremitting pruritus with sBA being permanently >250 μmol/L. At 18 years we proposed her to start a treatment with glycerol phenylbutyrate (GPB) (Ravicti, not labeled for the use under discussion) at the dose of 11 g/day (10 mL/day) of GPB ($8\,g \cdot m^{-2} \cdot day^{-1}$, in 2 divided doses) (5). GPB, a triglyceride consisting of 3 molecules of sodium phenylbutyrate joined to glycerol has no sodium burden, and offers palatability and pharmacokinetic advantages over PBA (5,6). Because of a better flavor and a convenient liquid presentation compared to PBA, full adherence to GPB treatment was obtained.

After 1 month, pruritus disappeared and sBA normalized (4.6 μmol/L). Follow-up at 3 and 8 months confirmed the absence of pruritus and cholestasis (sBA: 1.9 and 5 μmol/L). Serum liver tests, alpha-fetoprotein level, and liver elastography value (Supersonic Shear Imaging: 6 kPa) were normal. This report shows that in our PFIC2 patient GPB is as efficient as PBA to improve pruritus and cholestasis and allows good oral tolerance and therapeutic adherence.

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Telemedicine for Pediatric Inflammatory Bowel Disease in the Era of COVID-19

To the Editor: Telehealth is a widely adopted solution to maintain high-quality care for patients with chronic diseases while lessening the risk of transmission of SARS-CoV19 (1,2).

We would like to share our experience in expanding our telemedicine capability to address the comprehensive care needs for our pediatric inflammatory bowel disease (IBD) population.

Our Pediatric IBD center is located in the Bay Area, one of the earliest adopters of shelter in place. Given an existing telemedicine practice at our institution, we were able to convert appointments in less than a week to 100% telehealth visits. Our primary goals as we implemented our telemedicine program included the following:

1. Screen patients before infusion appointments, to keep infusion center safe.
2. Telehealth visits with patients receiving home infusion.
3. Injection teaching.
4. Routine care to our patients with IBD, including multidisciplinary visits (Pediatric Gastroenterologist, Nurse). Practitioner, Social Worker, Pediatric Dietician, Interpreter, and other specialists such as Surgery, Rheumatology, Immunology).
5. Provide urgent evaluations during flares to minimize emergency room visits and admissions.
6. Support ongoing IBD clinical trials.

We instituted a weekly virtual meeting with the care team to review acute issues, disseminate current literature on SARS-CoV-19 and IBD, and navigate limitations in available resources, such as nonurgent procedures. As we rethink our care algorithms to accommodate social distancing, we are also creating alternatives that we hope to continue beyond this pandemic (3).

Despite physical limitations we continue to promptly address questions, coordinate complex care, and triage clinical needs while enabling patients to stay at home, helping to reduce the spread of the virus to mass populations and the medical staff on the frontline.

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Heightened Central Line-associated Blood Stream Infection Risk During a Pandemic

To the Editor: We wish to update the pediatric gastroenterology community on a critical issue for children with intestinal failure (IF) that threatens to increase emergency department visits and hospitalizations during the pandemic related to