



phosphoethanolamine, consistent radiological features and was homozygous for a known TNSALP gene mutation. She had persistent hypercalcaemia, failure to thrive and at 3 months developed severe bronchiolitis requiring intubation and ventilation for 2 weeks. Following reports of dramatic improvements in mineralization for this previously untreatable disease in animal models<sup>1</sup> and humans,<sup>2</sup> asfotase alfa was commenced at 4 months (2 mg/kg subcutaneously (SC) three times per week). Biochemical disturbances resolved quickly and a further episode of bronchiolitis at 5.5 months was managed with nasal prong oxygen only; no further invasive ventilation has been required. She showed marked improvement of mineralization on serial radiographs, with straightening of her femori (Fig. 1). Her linear growth has been steady. She has been making excellent developmental progress, with no ongoing issues. She had persistent craniosynostosis, which required cranial advancement surgery at aged 3, which was uncomplicated with resultant normal cranial shape. Calcification was noted on radiography at injection site but nothing was palpable clinically – this had largely resolved on follow-up radiograph 1 year later with increased injection site rotation and switching to 1 mg/kg SC, six times per week. Nephrocalcinosis had been noted on ultrasound prior to commencement of ERT, but had resolved by age 4. This successful outcome is reflected in the clinical trials showing improved survival and respiratory outcomes,<sup>3</sup> with no significant side effects reported. We conclude that ERT should be strongly considered for this previously untreatable condition.

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Conflict of interest: PJ Simm and R Savarirayan are advisory board members for Alexion. R Savarirayan received speaker's fees from Alexion.

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Dear Editor,

## USE OF NICOTINE REPLACEMENT THERAPY IN YOUNG PEOPLE ENTERING CUSTODY IN NEW SOUTH WALES, AUSTRALIA

Haysom *et al.* found low rates of uptake and compliance with nicotine replacement therapy (NRT) among young people who smoke tobacco entering correctional facilities.<sup>1</sup> As the authors note, this is not surprising and reflects the findings of other studies.<sup>2</sup>

Management of nicotine dependence in adolescence is problematic. NRT has not been shown to be effective in this age group and bupropion and varenicline are not approved.

In the study by Haysom *et al.*, the fixed dose of nicotine used was probably inadequate for many smokers as only 14 mg or 7 mg patches were used. Combination NRT (nicotine patch combined with a quick acting product such as nicotine gum or lozenge) is considered best practice but was not available.

Other options, which are more acceptable to young smokers, are clearly needed to manage nicotine dependence in custody. One novel but controversial form therapy which is popular among young smokers is electronic cigarettes (e-cigarettes).<sup>3</sup> These battery-operated devices heat nicotine liquid into a vapour for inhalation and can alleviate cigarette cravings and nicotine withdrawal symptoms. Unlike nicotine patches, the dose of nicotine can be titrated to individual needs.

As well as delivering nicotine, e-cigarettes can satisfy the behavioural and sensory aspects of the smoking ritual. Furthermore, as there is no combustion, e-cigarettes are substantially safer than smoking tobacco.<sup>4</sup>

Special disposable e-cigarettes have been designed for high security environments and are being used in prisons in the UK and USA. They can provide long-term substitution for combustible cigarettes in custody and may help to prevent relapse to smoking after release.

E-cigarettes may be a more acceptable solution than existing treatments for managing nicotine dependence in young people in custody, especially when other treatments are refused or are not effective.

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Conflict of interest: CP Mendelsohn has received payments for teaching, consulting and conference expenses from Pfizer Australia, GlaxoSmithKline Australia and Johnson and Johnson Pacific. He has no financial or commercial relationship with any electronic cigarette or tobacco company.

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Dear Editor,

#### THE RACP EVOLVE GENERAL PAEDIATRICS LIST

*Evolve* is a joint initiative of the Royal Australasian College of Physicians (RACP) and its specialties to identify and reduce

low-value medical practices (tests, procedures or interventions that are overused, inappropriate or of limited effectiveness).<sup>1</sup> RACP specialties participate by producing a list of their 'top five' low-value practices to lay the ground for clinical change.<sup>2</sup>

In 2016, the RACP's Paediatrics and Child Health Division (PCHD) produced a top five list for general paediatrics. To kick-start the process, JS compiled a list of all paediatric-related clinical practices already identified as 'low value' by other RACP specialties and similar initiatives in Australia and overseas (<http://www.evolve.edu.au>; <http://www.choosingwisely.org>; <http://www.choosingwisely.org.au>; and <https://www.nice.org.uk>; accessed 24 May 2017). A core working group comprising six fellows (including HH and SD) and one advanced trainee discussed these practices and nominated others. JS conducted a rapid review of the published evidence to confirm that the practices were of low value, and 15 practices were shortlisted for further consideration.

**Table 1** Top 10 low-value paediatric clinical practices for all respondents according to sector

Ranking	All Do not routinely...	Public sector Do not routinely...	Private sector Do not routinely...
1	Prescribe oral antibiotics to children with fever without an identified bacterial infection	Prescribe oral antibiotics to children with fever without an identified bacterial infection	Advise frenotomy for the relief of ankyloglossia in newborns
2	Undertake chest X-rays for the diagnosis of bronchiolitis in children or routinely prescribe salbutamol or systemic corticosteroids to treat bronchiolitis in children	Treat GORD in infants with acid suppression therapy	Prescribe oral antibiotics to children with fever without an identified bacterial infection
3	Treat GORD in infants with acid suppression therapy	Undertake chest X-rays for the diagnosis of bronchiolitis in children or routinely prescribe salbutamol or systemic corticosteroids to treat bronchiolitis in children	Undertake chest X-rays for the diagnosis of bronchiolitis in children or routinely prescribe salbutamol or systemic corticosteroids to treat bronchiolitis in children
4	Order chest X-rays for the diagnosis of asthma in children	Order abdominal X-rays for the diagnosis of non-specific abdominal pain in children	Order chest X-rays for the diagnosis of asthma in children
5	Order abdominal X-rays for the diagnosis of non-specific abdominal pain in children	Order chest X-rays for the diagnosis of asthma in children	Order abdominal X-rays for the diagnosis of non-specific abdominal pain in children
6	Advise frenotomy for the relief of ankyloglossia in newborns	Advise frenotomy for the relief of ankyloglossia in newborns	Order baseline blood tests just because an intravenous cannula has been placed in a paediatric patient
7	Order baseline blood tests just because an intravenous cannula has been placed in a paediatric patient	Order baseline blood tests just because an intravenous cannula has been placed in a paediatric patient	Treat GORD in infants with acid suppression therapy
8	Undertake allergy testing such as skin prick tests or blood tests for the routine evaluation of eczema	undertake allergy testing such as skin prick tests or blood tests for the routine evaluation of eczema	undertake allergy testing such as skin prick tests or blood tests for the routine evaluation of eczema
9	Request an EEG after a child's first unexplained afebrile seizure	Request an EEG after a child's first unexplained afebrile seizure	Use SSRIs as the first-line intervention for mild to moderately depressed children and young people
10	Use SSRIs as the first-line intervention for mild to moderately depressed children and young people	Use SSRIs as the first-line intervention for mild to moderately depressed children and young people	Request an EEG after a child's first unexplained afebrile seizure

EEG, electroencephalogram; GORD, gastroesophageal reflux disease; SSRIs, selective serotonin reuptake inhibitors.