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### Problems involved with the use of comforters

While I share many of the concerns expressed by Gill’ in his diatribe on dummies there are a number of compelling reasons why the use of comforters should not be withdrawn. They work as a substitute amplification or correction. The first patent on the India rubber nipple resembling the present day dummy was recorded in 1845 and was described in use in its present form in London and Manchester in 1887. Unfortunately by the time the practice of dummyping the dummy in a variety of sweetening agents to make it a more effective pacifier had become established and this habit was noted to be associated with the early onset of dental caries. No doubt the loss of primary incisors mentioned by Gill is due to their destruction by rampant dental caries associated with the persistent use of sweetened pacifiers and their subsequent extraction due to spreading infection, pain, and loss of sleep. The association of dummy sucking with malocclusion is more complex than stated. While there is a general agreement on the effect of prolonged dummy sucking producing malocclusions in the primary dentition, these abnormalities are mainly self corrective on cessation of the habit which is usually before 5 years of age.

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### References

### Cataplexy in the Prader–Willi syndrome

We report cataplexy, sudden atomic episodes provoked by emotion, in three patients with Prader–Willi syndrome (PWS) and suggest that cataplexy may be relatively common in this condition.

Detailed questioning of the mother of an 18 year old woman who had PWS elicited a history of recurrent 30-min attacks of unconsciousness, apparent in death with laughter, with sudden loss of power in all the patient's limbs. If standing, she would slump to the floor but recover completely after a few seconds. She had no history of the sleep paralysis or hypnagogic hallucinations and there was no family history of cataplexy, narcolepsy, or epilepsy. Her EEG was unremarkable. Episodes of cataplexy and of narcolepsy, despite excellent weight control, have been reported by two other patients with PWS who attend this hospital, an 8 year old girl and a 10 year old boy. Only one of the three patients possesses the HLA DR15 (DR2) DQB1*0602 haplotype that is strongly associated with the narcolepsy–cataplexy syndrome.

Cataplexy is usually precipitated by emotion provoking laughter, anger, or joy. The affected individual often falls to the ground without losing consciousness and the phenomenon is often mistaken for an epileptic or cardiac event. It can occur in isolation as a dominantly inherited trait or in association with a number of other conditions (table 1). The association between PWS and cataplexy, though described previously,1,2 is not widely recognised. Suspected episodes of cataplexy have been reported in eight of 35 patients with PWS. However, cataplectic manifestations are often difficult to prove, requiring a detailed history that is perhaps seldom available or elicited. We suggest that cataplexy may be relatively common in PWS and enquires regarding its signs and symptoms should always be made, especially in any patient with a past diagnosis of paroxysmal events.

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**Table 1** Conditions in which cataplexy is a recognised feature

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>Familial isolated cataplexy</td>
<td>Narcolepsy syndrome</td>
</tr>
<tr>
<td>Norrie’s disease</td>
<td>Niemann-Pick disease type C</td>
</tr>
<tr>
<td>Offset-Lowry syndrome</td>
<td>Narcolepsy-cataplexy syndrome</td>
</tr>
<tr>
<td>Fontanemodified/hypothalamic structural lesions</td>
<td></td>
</tr>
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</table>
conjugated hyperbilirubinaemia, greatly in-finding no other similar cases, with a level of >1000. Liver investigations revealed similar findings to those previously reported, with an N-acetylglucosamine:creatinine ratio of 0.75.

A female infant, born to consanguineous Pakistani parents, was noted shortly after birth to have dysmorphic features, including prominent occiput, beaked nose, high arched palate, and arthrogryposis with dislocated hips and rocker bottom feet. Icthyosis was also present. Metabolic acidosis developed within a few hours of birth and severe conjugated hyperbilirubinaemia within two days.

Renal tubular acidosis was manifest by generalised aminoaciduria, phosphaturia, and an N-acetylglycosamine:creatine ratio of 0.75.

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Kawasaki disease following meningococcal septicaemia

A V Ramanan and E M Baildam

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