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Title: Gastrointestinal dysfunction in ventilated patients with Duchenne muscular dystrophy

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Body: Background: To evaluate mechanisms of abdominal discomfort in Duchenne muscular dystrophy patients with chronic respiratory failure we assessed symptoms and measured gastric-intestinal transport. Methods: In 33 DMD patients, age 9-38y, questionnaires, clinical examinations and the DMD impairment score (Kohler 2009) were obtained. Gastric and intestinal transits were measured by ¹³C breath tests and by radiography after ingestion of radio-opaque markers. Results: 23 of 33 patients complained of bloating and chronic constipation; 21 regularly used laxatives. 10 were not on PPV, 5 on PVV at night, 8 on 24h PPV. Gastric emptying and intestinal transit were increasingly delayed with older age and higher impairment scores.

Gastro-intestinal function in DMD patients

	Age, y	DMD physical impairment score	Gastric emptying, min	Intestinal transit, h	Colonic transit, h	Constipation present
No PVV, n=10	19.0±6.8	51±14	219±58	6.9±1.9	63.4±19.3	3 of 10
On PPV, n=23	25.9±5.8*	66±6*	187±60	6.7±2.6	65.5±29.9	19 of 23*

Means±SD. *P<0.05 vs. no PPV. DMD impairment scores range: 10 to 80 with increasing impairment. Constipation was rated subjectively as present or absent. Intestinal transit norm < 5h (Geypens 1999); Colonic transit norm < 50h (Southwell 2009)

Use of PPV was correlated with accelerated gastric emptying when controlled for impairment (R=-0.43, P=0.048). Massive gastro-intestinal air accumulation was common. Conclusions: Gastro-intestinal disturbances in DMD patients are common and related to delayed gastric and intestinal transit with

increasing age, progression of physical impairment and respiratory failure. PPV may accelerate the delayed gastric emptying, possibly by esophago-gastric air inflation.						