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MRI assessment of left ventricular structure and function in children with infantile Pompe disease

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Introduction

Pompe Disease (α-glucosidase deficiency, glycogen storage disease type II) is a progressive disease resulting from accumulation of lysosomal glycogen in skeletal and myocardial cells. In the infantile form there is rapid progression of disease and rare survival beyond the first year. Severe left ventricular (LV) hypertrophy is characteristic but may regress with enzyme replacement therapy (ERT). Cardiac MRI (CMR) is useful to follow LV mass, but sedation is especially high risk. We report our success in performing CMR in this population with minimal or no sedation.

Methods

CMR was performed using a 1.5 Tesla scanner. Non-ECG gated True-FISP real-time imaging, using an imaging matrix of 90 × 128, 200 × 140 cm FOV, 5 mm slice thickness with 5 mm gap and 80 degree flip angle, resulted in a cine image with a temporal resolution of 55-60 msec. Retrospectively ECG gated True-FISP cines acquired during free-breathing with multiple averages and an imaging matrix of 92 × 128, 200 × 140 cm FOV, 5 mm slice thickness with 5 mm gap and 80 degree flip angle resulted in a temporal resolution of 30 msec.

Delayed enhancement was evaluated using either inversion recovery True-FISP single shot or segmented Turbo-Flash sequences in short axis and long axis views 5-10 minutes after IV administration of 0.2 mMol/kg of gadolinium. The inversion time was set to null normal myocardium.

LV mass and function were measured from the interpolation of endocardial and epicardial contours from a stack of short axis slices, with inclusion of the trabeculations and mitral papillary muscles. Results were normalized to body surface area.

Results

Study results are presented in Table 1. 10 subjects underwent a total of 17 CMR studies. 13 CMR studies were performed without sedation, while sedation supervised by pediatric cardiac anesthesia was provided for 4 studies. The median LV mass by CMR was 135 gm/m^2 (range 34-334 gm/m^2) with a median LV ejection fraction of 57% (range 18-73%). In 5 subjects with serial CMRs, there was a trend towards reduced LV mass with ERT, although one patient had an increase in LV mass despite ERT. Of all the 17 CMR studies, only 1 demonstrated delayed enhancement.

Conclusion

CMR is useful to longitudinally follow children with Pompe disease and their myocardial response to ERT. CMR can be performed safely without sedation in this high risk population. In contrast to other glycogen storage diseases, children with Pompe disease appear to have less myocardial scar.

Table 1:

Subject	Sex	Age(m) at start of ERT	Months post-ERT	MRI Sedation	MRI Technique	LVM (gm/m2)	LV EF	Delayed Enhancement
1	F	4	7	No	Cine-TrueFISP	135	25%	No
1			9	No	Real-Time	180.6	58%	No
1			13	No	Real-Time	179.6	55%	No
2	M	7	15	No	Real-Time	51.2	55%	No
2			26	No	Real-Time	57	57%	No
2			47	Yes	Both	44.5	68%	No
3	M	6	1	No	Real-Time	334	18%	No
3			2	No	Real-Time	303	25%	No
4	M	3	6	No	Real-Time	43.8	55%	No
4			36	Yes	Real-Time	43	69%	No
5	M	1	0	No	Real-Time	94	73%	No
5			20	Yes	Both	34	57%	No
6	M	4	34	No	Cine-TrueFISP	47.0	60%	No
7	M	6	2	No	Cine-TrueFISP	252.2	65%	No
8	M	4	3	No	Real-Time	180.6	29%	No
9	F	10	1	No	Real-Time	324.4	55%	No
10	M	3	31	Yes	Both	145.0	73%	Yes

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