

QA 42 - Wolf Hirschhorn Syndrome

QUESTION:

I am seeing a 7 month old girl with Wolf Hirschhorn Syndrome. She was born at 39 weeks gestation at 3 lbs. 7 oz. At 6 months, she weighed 7 lbs. 13 oz. and was 21 ¼” long. She had a video swallow study and was having some aspiration on plain formula, but was okay when the formula was thickened. Her mother states that she takes 24 oz. per day of Enfamil thickened with 2 tablespoons of cereal per 4 oz. of formula. This calculates to 170 calories per kg. She has a small cardiac defect, but the cardiologist does not think it is interfering with her growth.

Is there information about what kind of growth we should expect for this child? I keep talking to her mother about concentrating the calories in the formula even further, but her physician’s don’t see that as a priority.

ANSWER:

Battaglia, et al described 15 cases of Wolf-Hirschhorn Syndrome from infancy to 16 years at the time of publication. All patients had short stature and slow weight gain with “adequate” calorie and protein intake, whether or not they had cardiac defects. They do not comment on weight for length ratio. Patients all had hypotonia and muscle hypotrophy and had lower than normal weight for any given length. Thus a weight/length ratio between the 5-10th percentiles may be acceptable. In seven cases with feeding problems, a gastrostomy tube placement was needed. An interesting finding in this article was that patients tended to improve developmentally over time. Three patients eventually were able to walk, unsupported, and two were able to walk with support. Abilities to communicate and to perform some self help skills improved in later childhood years.

This article implies that it is important to deal with issues such as failure to thrive, in order to optimize development over time. The patient you describe has gained an average of 9 grams/day over the first 6 months of life, while “normal” (50th percentile) weight gain would be an average of 21.5 grams/day and “low” (5th percentile weight gain) would be 14.5 grams/day. Her SGA birthweight may also impact expected growth in addition to the diagnosis. It would be important to verify whether or not your infant is taking the full 24 ounces/day of formula reported by her mother. It would be reasonable to observe feeding to see how much formula is actually ingested from a bottle, and to have mother keep a written record of feedings every day for a week. If feeding is very difficult, and/or inefficient, and/or the baby is taking in much less than originally reported, it may be necessary to consider naso-gastric or gastrostomy feedings.

References:

- 1) Battaglia, A., Carey, J., Cederholm, P., et al Natural History of Wolf-Hirschhorn Syndrome: Experience with 15 Cases. *Pediatrics* 1999; 103:830-836.
- 2) Guo, Shumel, Roche, Alex, Fomon, Samuel, et al, Reference Data on Gains in Weight and Length during the First Two Years of Life. *J Pediatr* 1991; 119: 355-62.