

Cyclic Vomiting Syndrome Association USA/Canada

3585 Cedar Hill Rd. NW, Canal Winchester, OH 43110

Telephone 614/837-2586 Fax 614/837-2586

Email waitesd@cvsaonline.org

Website <http://www.cvsaonline.org>



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Uretero Pelvic Junction Obstruction Discovered - Mimics CVS

An International E-Mail Discussion of Importance

Kathleen Adams

Research Liaison, CVSA

The following message came to me recently from a CVSA mom whose child was a participant in Dr. Boles' study on mitochondrial disease with CVS. I forwarded the message on to the professional email distribution list. The vigorous professional discussion follows.

Dear Kathleen,

Thank you for the update, however our family will not be needing anymore information from C.V.S.A. Our child became extremely ill in June and was hospitalized twice in one month. While our pediatrician saw no need to review his case and pushed for a premature discharge, my dad insisted on a surgical consult. It was an ultrasound that immediately revealed a U.P.J. obstruction and a kidney that was 7x's the size it was 2 1/2 years ago. I was very concerned with his high doses of Elavil (amitriptyline) and his worsening condition. He has now had his operation and stint removal. Surgeons were shocked that his kidney still functioned and will continue to check him every 3 months. We praise God for this resolution to our continual suffering and we are thrilled that despite the damage, his kidney is alright. Thank you for all you have done to help our family.

TO: Professional CVSA distribution list

From: Kathleen Adams

I just wanted to reinforce this differential diagnosis clue for sorting out the cause of cyclic vomiting.

CVSA. This message is from one of the families in the control

study for Rick Boles, MD mitochondrial/cvs study: "Search for mitochondrial DNA mutations in individuals with CVS". She emailed back in response to my update on study progress.

Schulte-Bockholt A, Werlin S et al: *Ureteropelvic junctions obstruction: an overlooked cause of cyclic vomiting*. American Journal of Gastroenterology 97(4):1043-1045, 2002

From: a physician

What is the best way to assess for UPJ obstruction? Should we do it in every CVS patient?

Kathleen's response:

I'm going out on a non-diagnostic limb. My opinion comes from memory from David Fleisher's & B Li's lectures about using ultrasound, & a keen eye. Dr. Fleisher has a slide that shows a frontal view of child with very subtle bulge on left flank who is later found out to have a UPJ obstruction. I think clinicians would agree that it should be screened for on every patient.

From Dr. Jean Perrault, MD - Peds GI - Montreal Children's Hospital

Thanks for the reminder. It is interesting that it should come one day after we picked up such a patient. In this instance, the first study, while the patient was asymptomatic, was negative, but we did instruct the parents to make sure to return during an attack so that we could repeat the study (while trying prophylaxis with erythromycin) and indeed the repeat study was now quite positive!

From: Larry Adams - DVM - father to 2 children with CVS, CVSA Board of Directors member

This is an important differential for CVS and it should not be ruled out by only one ultrasound exam while the child is normal. UPJ obstructions can be intermittent and resolve spontaneously, then recur at variable intervals; therefore the signs are very similar to CVS. The best time to do the ultrasound is while the child has symptoms and/or to repeat the study at least once with careful comparison back to the initial evaluation. My MD urology colleagues (whom I work with for lithotripsy of kidney stones) were impressed how similar the clinical presentation of CVS is to what they see as classic intermittent UPJ obstruction.

From Richard G. Boles, M.D. - Medical Genetics, Childrens Hospital Los Angeles

This underscores Dr. B Li's recommendations for all CVS patients to have a radiological (x-ray or ultrasound) test of the abdomen. I am glad that the doctors caught this in time.

From Gill McDonald - Board of Trustees, CVSA UK

One of our UK members, originally diagnosed with CVS, also had to have kidney surgery, I think because of the same obstruction. Recently one of our former committee members with a child diagnosed with severe CVS found that the trouble was in fact a missed malrotation of the bowel needing urgent surgery. This was not discovered until the parents insisted the child was referred to our medical advisor, Dr. Sonny Chong. He spotted it and referred her to a surgeon who operated immediately.

Barry Lipschitz, MD - Paediatric Gastroenterologist, Melbourne, Australia

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I was interested to read about this case of UPJ obstruction. I recently discovered a similar situation in a boy who I thought had Abdominal Migraine. I had an abdominal ultrasound done due to suspicion of a renal problem and found evidence of an enlarged kidney which is probably partially obstructed. I don't yet know the pathology - he is pending surgery. Thanks for the reference.

From: George Russell - Emeritus Professor, University of Aberdeen, Scotland

I do not wish to appear critical in any way of this child's management, but I do feel the family's problems resulted at least in part from a failure to understand the nature of cyclic vomiting syndrome. Cyclic vomiting syndrome is just what it claims to be, ie a syndrome, and a syndrome is nothing more than a collection of symptoms and signs that occur together in a way that suggests that they are related. Over time a syndrome may turn out to be a single disease (ie a single pathological process), but in other cases the syndrome may turn out to be the end-point of a number of different diseases. It has always been clear that the clinical syndrome of cyclical vomiting may be due to a variety of different diseases, such as disorders of intermediary metabolism and urinary tract problems. Ishaq Abu-Arafeh and I wrote a chapter on migraine-related disorders in his new book (I Abu-Arafeh, ed. *Childhood Headache*. Cambridge University Press, 2002, ISBN 1-898-68326-3). After discussing the diagnostic criteria for cyclic vomiting syndrome, we went on to say:

Nevertheless, despite the undoubted value of these criteria, CVS is not a diagnosis to be made lightly. There are numerous causes of recurrent vomiting in children, including-

- intermittent intestinal obstruction secondary to intestinal duplication or stenosis, mid-gut malrotation and other lesions
- urinary tract disease including infective and obstructive lesions
- raised intracranial pressure
- endocrine disorders such as Addison's disease
- peptic ulceration
- inborn errors of metabolism, particularly urea cycle disorders and intermittent forms of short chain organic acidaemias

It is essential therefore, even when the above diagnostic criteria are met, that these children are investigated thoroughly, particularly if they are encountered at an early stage of their illness, before periodicity and interval wellness are properly established, or if there is any suggestion of chronicity of the symptoms. The diagnostic work-up should include-

- urine culture
- biochemical screen
- abdominal ultrasound, looking particularly for renal enlargement
- urine examination for amino-acids, short chain organic acids and porphyrins during an attack (urinary ketones are normally present during an attack, giving rise to acidosis as an alternative name for the syndrome)
- acid-base balance, lactate/pyruvate and ammonia during an attack. This list is by no means exhaustive, and may need to be extended if there are hints of chronicity.

We should therefore continue to use the term 'syndrome' in describing cyclic vomiting, as a constant reminder that we are not dealing with a single well-established disease. Never mind the cost-benefit analysis, CVS is a horrendous condition, and we owe it to our patients to make sure there is no underlying anatomical or biochemical disorder.

From Dr. B.U.K. Li, MD - CVS Program- Children's Memorial Hospital, Chicago, IL

In all of my recent talks, I speak very strongly about excluding hydronephrosis with UPJ obstruction indicating that we have seen 10 such cases - most frequent surgical lesion. YES, this is important. We do a study when well because of scheduling (this will pick up most according to the urologists) and occasionally repeat when ill, if still concerned.

From Ramon Tormo, MD - Head Unit of Paediatric Gastroenterology Unit, Hospital "Vall d' Hebron", Barcelona, Spain.

Your case is very interesting, but usually this diagnosis can be ruled out...more difficult are the cases of BETA-OXIDATION DISEASES, or HETEROZYGOTES of UREA CYCLE DISEASES.

From David Fleisher, MD - Peds GI, University of Missouri - Columbia

Dear Dr. Russell: I couldn't agree more with your admonition for open-minded approach to diagnosis of CVS!

From Ernesto Guiraldes, MD

Chief Peds GI Group, Pontificia Universidad Catolica
Santiago, Chili

Professor's Russell reply is a sensible and welcome reminder of what CVS really is. It is a highly enlightening comment. Thank you very much.

From Sonny Chong, MD - Consultant Paediatrician, GI, Queen Mary's Hospital for Children, Surrey, UK

It is good to hear so many replies including Dr. George Russell's very sage comments. At our last CVSA UK family day in November at Birmingham Children's Hospital, we revisited this very issue - that it is still very much a syndrome, rather than the view that CVS was synonymous to migraine

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and little else. In the last three years in the UK, intestinal migraine had to be turned on its head twice when our patients came back with a diagnosis of malrotation after being misdiagnosed (Gill McRonald had mentioned earlier), and another which turned out to have hereditary familial pancreatitis 10 years later as an adult.. We need to keep an open mind, and not pigeon hole patients, from which it becomes hard to unravel the true etiology. Regards

From Prof. David CA Candy, - Paediatric Gastro-Enterology & Nutrition Service Royal West Sussex NHS Trust Chichester, UK. The advice from our Paediatric Nephrologist was that a normal renal scan between attacks of vomiting does not exclude PUJ obstruction and the scan must be performed during obstruction and the scan must be performed during an attack. This can take some persuading at 3:00 am! More data would be welcome.

From Gunnar Stickler, MD - Emeritus Professor Pediatrics, Mayo Clinic and Mayo Medical School, Rochester, Minnesota

In 1971, we reported on 109 children with ureteropelvic obstruction. I quote from this study: "in a third of the children the predominant symptom was vague abdominal pain in repeated attacks, frequently localized around the umbilicus and associated with cyclical vomiting." Kelalis,Culp,Stickler and Burke: "Ureteropelvic obstruction in children: experiences with 109 cases" *J.OF UROLOGY* (1971)106:418 Gunnar B. Stickler

From Alberto Ravelli, MD - Peds GI, Univ of Brescia - Italy

Although identification of a minor UPJ obstruction can be challenging, I share Dr Tormo's view that the most difficult "known" causes of CVS to be ruled out are metabolic disorders such as urea cycle and beta-oxidation defects. As usual in medicine, however, every disorder can be easy (or difficult) to identify if you think (or do not think) about it. Regards

From Dr David N.K. Symon - Consultant Paediatrician, University Hospital of Hartlepool UK

PUJ obstruction (as it is called in Britain) is well known as a differential diagnosis of CVS. David Forbes mentions it in his paper on differential diagnosis in the proceedings of the first symposium in London, and it is in my book chapter. The danger of diagnostic criteria is that they are used as a "Tick List". If you have ticks in all the boxes you must have the condition. It doesn't work. Symptoms of CVS are the final pathway of several disease processes. Any research which lumps together all patients with CVS as if they have a single condition is likely to be inconclusive, as we are dealing with more than one thing. ♦

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Cyclic Vomiting Syndrome Association-USA/Canada, founded in 1993 actively pursues the following mission:

"CVSA USA/Canada will raise awareness, provide education and support to those affected by CVS and advocate for research about nausea and vomiting."