

## Celiac Artery Compression in Children, Adolescents, and Young Adults

**To the Editor:** The article by Scholbach<sup>1</sup> on celiac artery compression syndrome is interesting in that it evaluates patients from 0 to 18 years. I note that he identifies a prevalence of sonographic criteria of celiac artery compression syndrome of 1.7% in this age range. In our study,<sup>2</sup> although we did not evaluate patients specifically during childhood and adolescence, we found that celiac artery stenosis was common in patients older than 65 years (18% of patients) when it was usually associated with atheromatous disease but was also found in up to 3% of patients younger than 65 years. All of our patients were asymptomatic and did not demonstrate any of the symptoms that Scholbach's report alludes to. It is important to recognize that celiac artery abnormalities can occur in the absence of symptoms if inappropriate treatment is not to be instituted. Careful evaluation of the symptoms and their relationship to potential perfusion abnormalities is important before undertaking surgical or minimally invasive intervention.

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### Reply

**To the Editor:** I gratefully take the opportunity of Dr Dubbins' letter to go into more detail about the issue of correlation of celiac artery compression as diagnosed by color Doppler sonography and possibly related clinical symptoms.

My article<sup>1</sup> points primarily to the sonographic observation of a constellation in young patients that is known as a distinct entity in adults so far. I attempted to show that children may have sonographically relevant compression of the celiac artery, and many of them in this study had symptoms that seemed unusual in this situation and could not be explained by other findings.

It is obvious that this alone is not proof of any causal relationship between the sonographic observation per se and such symptoms. The relief of symptoms after surgical correction would be the most sound proof. Because of lack of knowledge about this disorder in preadult patients, criteria for when to operate and on

whom must be elaborated. The goal of my article, therefore, was to focus the attention on celiac artery compression. It is interesting, however, to rethink pathophysiologic hypotheses that try to link both sonographic and clinical symptoms.

My impression is that pure hemodynamic approaches to the problem fall short. It is barely imaginable that malperfusion of the liver, stomach, and spleen should evoke severe (hypoxic?) pain in these adolescents. In fact, I did not find collateral vessels in most of my patients. This should be expected as an adaptive response to hypoperfusion or hypoxia. It is more likely that pain is evoked by irritation of the celiac ganglion. It is well documented that irritation of this ganglion may cause severe pain attacks.<sup>3,4</sup> The function of the celiac ganglion is to coordinate vegetative processes in the abdomen. My observation is that vegetative symptoms are common in patients with celiac artery compression, and in some patients, just these vegetative symptoms (along with pain in many) can be provoked by a 2- to 5-second single-finger compression of the celiac artery's origin. I vividly recall some patients with flush, nausea, pain, and vertigo provoked by this maneuver. I did not mention this in my article because I did not investigate this systematically. Altogether, these are hints to a major role of the celiac ganglion in "celiac artery compression syndrome" or "arcuate ligament syndrome." The latter term does not restrict thoughts to the artery alone. Despite this, I preferred "celiac artery compression" to point to the sonographically observable arterial compression. Unfortunately, as far as I know, we cannot detect the ganglion by means of sonography.

Dr Dubbins refers to his observations in 184 patients 4 to 92 years of age (details on ages were only given in a diagram of his article; only 4 seemed to be younger than 18 years).<sup>2</sup> In fact, his article deals with a different entity, celiac artery stenosis, whereas I referred to celiac artery compression. This seemingly tiny difference may turn out to be relevant from a pathogenetic point of view. Compression from outside will inescapably affect surrounding structures, the celiac ganglion being by far the most important. This cannot be assumed in a case of an internal stenotic, atheromatous narrowing of the

vessel. Thus, it is only logical that symptoms may discriminate both diseases. Vegetative dysregulation mediated by an irritated celiac ganglion must be missed in cases with internal diameter reduction due to arteriosclerotic changes. Effects on hemodynamics are also quite different: constant reduction of flow volume in arteriosclerotic disease of the artery versus intermittent reduction in celiac artery compression. Therefore, it might not be surprising that there are differences in symptoms in both entities.

Nevertheless, I also made the observation that a portion (8 of 59 patients) were free of symptoms (14%) at the time of their investigation (all pain sensations were contracted wherever localized):

No. of Symptoms	%
0	14
1	22
2	24
3	20
4	12
5	5
6	3

In prospective investigations, the rate of asymptomatic patients is expected to be higher. Therefore, the observation of celiac artery compression is a challenge for all of us dealing with young patients with vegetative and pain symptoms to contribute our experiences. Reports of patients who have had surgery would be most valuable. Here, it seems wise to exclude relevant differential diagnoses and to draw on indications as in adult patients initially: severe pain attacks or weight loss. In patients with additional vegetative symptoms, their postoperative course should be carefully followed. This could give a hint if it someday could be justified to operate on patients solely with vegetative symptoms. These kinds of symptoms may be very pronounced and may be a strain for some patients.

I personally know of 5 teenagers who had successful surgery with prompt and lasting pain relief: 1 underwent open surgery, which was carried out in a neighboring clinic,<sup>5</sup> and 4 had minimally invasive surgery at another hospital (Annegret Klimas, MD, personal communication, 2006). After submission of my paper, I became aware of a large series of Russian adolescents who had successful operations.<sup>6</sup> The authors described neurovegetative symptoms

and pain as indications for operating on celiac artery compression and relied mainly on color Doppler sonography. Their success rate was 90% (87/97).

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