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**Review Article** 

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# EVOLUTION OF SUPERIOR MESENTERIC ARTERY SYNDROME: A HISTORICAL PERSPECTIVE

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## INTRODUCTION

Superior Mesenteric Artery syndrome, also known as Wilkie's syndrome or cast syndrome, is a rare condition resulting from vascular compression of the duodenum. Its historical journey reflects the broader evolution of gastroenterology, surgery, and diagnostic imaging. Early descriptions of SMA syndrome were anecdotal, but the advent of modern medical tools has significantly expanded its understanding. This review aims to trace the history of SMA syndrome, from its initial recognition to present-day management, to contextualize its place in medical science.

#### Historical background

The origins of SMA syndrome date back to the late 19th century. The condition was first described by Carl Freiherr von Rokitansky in 1861, who observed compression of the duodenum by the aorta and superior mesenteric artery during autopsy studies.<sup>[1]</sup> However, it was not until 1927 that Austrian surgeon David Wilkie, Carl von Rokitansky's successor, published a comprehensive series of 75 cases, solidifying the syndrome's clinical recognition.<sup>[2]</sup> This landmark paper provided detailed insights into the anatomical and clinical features of the condition, laying the groundwork for subsequent research.

Beginning in the 1840s, various terms have been used for over a century to describe the phenomenon of proximal intestinal obstruction and dilation caused by compression of the third portion of the duodenum by the superior mesenteric artery (SMA), or more specifically, between the SMA and the aorta. Carl von Rokitansky (1804– 1878) initially referred to this condition under the name "internal hernias," which he associated with incarceration.<sup>[3]</sup> Nearly 20 years later, he redefined the condition as an "internal incarceration" or "intestinal incarceration". He described it more thoroughly as follows:

"The incarceration is due to the pressure which a portion of the intestine and its mesentery exert on a piece of intestine, bearing on it and compressing it from the front to the rigid posterior abdominal wall. Here, compression of the lower transverse part of the duodenum is caused by the small intestine mesentery, namely the mesenteric

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artery, which enters the superior mesenteric root with the surrounding nerve plexus, compressing the S-loop or the last portion of the ileum by a downward displacement of the small intestine".<sup>[4]</sup>

In 1908, in his paper "Acute Dilation of the Stomach and Anterior-Mesenteric Ileus," Laffer drew attention to the variety of terms used to describe this clinical entity, including: acute dilation of the stomach, arteriomesenteric ileus, gastro-mesenteric ileus, mesenteric intestinal obstruction, post-operative ileus, post-operative arterio-mesenteric intestinal obstruction, post-operative acute dilation of the stomach, acute duodeno-jejunal intestinal obstruction, post-operative gastric paralysis, combined ileus, duodenal ileus, duodenal and compression.<sup>[5]</sup>

Other terms used included acute gastroduodenal obstruction and mesenteric duodenal compression. It was David Wilkie (1882–1938) who referred to the condition as chronic duodenal obstruction in 1921.<sup>[2]</sup> The eponym Wilkie's (or Wilke's) syndrome, or duodenal ileus arterio-mesenteric ileus, was first used by Grauer in 1948 to honor Wilkie's accomplishment in providing the first comprehensive description of this disease in 75 patients in 1927.<sup>[6,7]</sup> The term *Cast syndrome* was introduced by Dorph in 1950 to describe signs and symptoms caused by abdominal compression from a hip spica cast or fullbody cast.<sup>[8]</sup> Finally, the term *SMA syndrome* was adopted by Kaiser et al. in 1960.

#### Advancements in diagnostic understanding

*Early diagnostic challenges:* Before the advent of modern imaging, SMA syndrome was primarily diagnosed based on clinical symptoms and exploratory laparotomy findings. Patients often presented with symptoms of intermittent postprandial pain, nausea, vomiting, and weight loss, which were frequently mistaken for more common gastrointestinal disorders.

*Introduction of radiological imaging:* The mid-20th century marked a significant turning point with the introduction of radiographic techniques. Barium meal studies became the standard diagnostic tool, enabling visualization of the characteristic duodenal compression.<sup>[9]</sup> In the 1970s, computed tomography (CT) and angiography further enhanced diagnostic accuracy by providing detailed anatomical views of the vascular structures.<sup>[10]</sup>

*Modern diagnostic tools:* Recent advancements, such as three-dimensional CT angiography and magnetic resonance imaging (MRI), have revolutionized the understanding of SMA syndrome. These modalities allow for precise measurement of the aortomesenteric angle and distance, key parameters in diagnosing the condition.<sup>[11]</sup> Endoscopic evaluation has also emerged as a valuable adjunct in assessing duodenal obstruction.

#### **Evolution of treatment approaches**

*Conservative management:* Historically, conservative treatment focused on nutritional rehabilitation to restore retroperitoneal fat and widen the aortomesenteric angle. This approach gained prominence in the mid-20th century, supported by studies demonstrating symptom resolution in patients following weight gain and nutritional therapy.<sup>[12]</sup>

*Surgical interventions:* Surgical treatment, initially limited to gastrojejunostomy, evolved significantly over time. The advent of laparoscopic and robotic-assisted techniques has made duodenojejunostomy the preferred surgical approach, offering minimally invasive solutions with reduced morbidity.<sup>[13]</sup> Other innovative procedures, such as Strong's procedure and Roux-en-Y reconstruction, have been explored for refractory cases.

*Emerging therapies:* In recent years, multidisciplinary approaches incorporating physical therapy and psychological support have been emphasized. Emerging therapies, including endovascular stenting and bioengineering techniques to modify vascular anatomy, are under investigation.<sup>[14]</sup>

### **Controversies and Challenges**

Despite advancements, SMA syndrome remains a subject of debate. Critics argue that its rarity and nonspecific symptoms complicate diagnosis, often leading to overdiagnosis or misdiagnosis. Furthermore, the condition's exact prevalence and pathophysiology are still not fully understood. Efforts to standardize diagnostic criteria and treatment algorithms are ongoing.

#### Impact of historical context

Understanding the historical evolution of SMA syndrome provides valuable insights into the interplay between medical advancements and clinical practice. The recognition of this condition highlights the importance of anatomical studies, while the refinement of diagnostic tools underscores the role of technological innovation in shaping medical knowledge. The challenges faced by early clinicians in diagnosing and managing SMA syndrome continue to inform contemporary strategies.

#### CONCLUSION

The history of Superior Mesenteric Artery syndrome exemplifies the dynamic nature of medical science. From its initial description in the 19th century to current multidisciplinary approaches, the journey of SMA syndrome reflects the continuous pursuit of knowledge and innovation. By appreciating its historical development, clinicians and researchers can better address the challenges posed by this enigmatic condition and foster advancements in its diagnosis and treatment.

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