Currarino Syndrome: A Rare Case Report of Sepsis and Its Management

Polyxeni Theodosopoulou*, Georgia Micha†, Ioannis Koutalas‡, Stavroula Karachanidi§, Panagiota Brattou¶, Anteia Paraskeva**

Currarino syndrome (CS) is characterized by a triad of anomalies consisting of: a sacral bone defect, anorectal malformations and a pre-sacral mass. We present the case of an adult patient with a medical history of CS who presented with septic shock and was subjected to an emergency laparotomy due to severe abdominal distension. In this particular case, we underline the importance of immediate surgery on the patient's outcome as well as the considerable role of landiolol in controlling the heart rate with no further deterioration of blood pressure in this patient presenting with atrial fibrillation and sepsis. [*P R Health Sci J 2022;41(3):168-171*]

Key words: Currarino syndrome, Atrial fibrillation, Septic shock, Landiolol, ICU

Currarino syndrome (CS) is an autosomal dominant hereditary disease characterized by a triad of abnormalities, comprising of partial sacral agenesis, anorectal malformation, and a pre-sacral mass (anterior meningocele, enteric cyst, or teratoma) (1). Patients with CS usually suffer from prolonged constipation, which is at times, the only clinical presentation. A possible mechanism of such a defecation disorder is the anterior position of the anus or the compression of spinal nerve roots due to a pre-sacral mass (2). These temporary disturbances of gastrointestinal motility can lead to long-term constipation, chronic megacolon and paralytic ileus (3). We present an overview of the acute-sepsis management of an adult patient with CS and atrial fibrillation (AF).

Case Report

We report the case of a 53-year-old male patient with a history of CS (since childhood) and with concomitant systematic scleroderma, arterial hypertension, and episodes of AF;, who presented to the emergency department with acute sepsis state and severe abdominal distension. Both approval and written informed consent were solicited and received from the patient.

The patient was tachypneic, suffering from hemodynamic instability and metabolic acidosis (pH 7.05), and meeting 2 of the quick Sequential Organ Failure Assessment (qSOFA) criteria (high respiratory rate>22 breaths per minute and low systolic blood pressure<100 mm Hg) as well as 2 of the SOFA criteria (impaired renal status and the need for continuous noradrenaline infusion). Following the 2016 consensus guidelines for sepsis, the above, along with the patient's over-distended abdomen and his history of CS, helped establish the diagnosis of sepsis, making the patient a candidate for an emergency laparotomy. The sigmoid was proven to be distended to a diameter of 25 cm, raising intra-abdominal pressure and compromising our patient's ventilation. Pressure- controlled mechanical ventilation was preferred at first,; however, after exploration manoeuveres and suctioning, the respiratory mechanics improved.

Intraoperatively our patient's cardiac rhythm ranged from sinus tachycardia to atrial flutter and AF. During sepsis, the management of tachycardia accompanied by hypotension can be challenging. While direct current cardioversion is a guideline recommended treatment for acute onset AF in unstable patients, it is often unsuccessful for rate control during sepsis (4). B-blockers and calcium blockers may slow atrioventricular nodal conduction and lower heart rate but can also worsen hypotension in patients who are in a state of distributive shock (such as septic shock) (5). A highly selective b1-blocker, Landiolol, was favoured over other similar intravenous agents in order to achieve a heart rate control of less than 110 bpm with the least possible haemodynamic instability since our patient was already on 2 vassopressors (noradrenaline and arginine vasopressin) and hydrocortisone infusion in order to sustain an adequate mean arterial pressure (MAP> 75 mm Hg). The need for slowing the ventricular response in a context of severe hemodynamic instability was very well achieved through continuous Landiolol infusion (along with the other vasopressors). Not only was the heart rate reduced

*Resident, Department of Anesthesiology, Aretaieion University Hospital, Athens, Greece; †Anesthesiologist, MD, PhD, Department of Anesthesiology, Elena Venizelou Hospital, Athens, Greece; ‡Anesthesiologist, Aretaieion University Hospital, Athens, Greece; §Resident, Department of Anesthesiology, Aretaieion University Hospital, Athens, Greece; ¶Resident, Department of Anesthesiology, Aretaieion University Hospital, Athens, Greece; **Assistant Professor of Anesthesiology, MD, PhD, DESA, Department of Anesthesiology, Aretaieion University Hospital, Athens, Greece

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Address correspondence to: Polyxeni Theodosopoulou, 34 Antifilou St, 11528, Athens, Greece. Email: xeniathd@gmail.com

by a significant amount, but hemodynamic variables such as blood pressure managed to remained stable. (Table 1).

The procedure lasted 3 hours, at the end of which, the patient was transferred, intubated and sedated, back to the ICU. The surgical technique performed was a Hartmann sigmoidectomy with the formation of an end colostomy. During the surgical manipulations, a large pre-sacral mass compatible with the patient's history of CS was encountered. The mass was a myelomeningocele. After a successful weaning process, he was extubated on the first postoperative day, and noradrenaline and arginine vasopressin were subsequently discontinued. Landiolol was switched to esmolol on the second post-operative day, since the patient was hemodynamically stable. He was released from the hospital on the 10th post-operative day, hemodynamically stable and afebrile.

Table 1. Intraoperative values of our patient's blood pressure and heart rate before and after the beginning of landiolol infusion.

	Blood Pressure (mm HG)	Heart Rate (bpm)
Before landiolol infusion (with constant infusion of 2 vasopressors)	120–130/45–55	120–130
After landiolol infusion (with constant infusion of 2 vasopressors)	120–140/55–65	90–110

Discussion

Firstly described as a syndrome in 1981, CS is defined by a triad of anomalies consisting a sacral bone defect, anorectal malformations and a pre-sacral mass; it represents only 5% of all symptomatic anorectal malformations. Bowel obstruction (in infancy), intractable constipation, urinary retention, incontinence, and infection are symptoms that are often associated with this condition (6). An adult diagnosis of CS is relatively unusual. In our patient, the anterior position of the anus and the pre-sacral mass compression of sacral nerves (nerves that innervate the distal colon) contributed to decreased rectal tone and motor function. Furthermore, these episodes of the transient inhibition of gastrointestinal motility caused long-term constipation, chronic megacolon, and, finally, paralytic ileus in the patient.

Bowel obstructions in patients with CS have, when present, been variously attributed to rectal malformation-associated obstruction, spinal cord tethering and obstruction from the pre-sacral mass. (1). However, none of these explanations adequately accounts for the presence of such a severe distension of the colon. Our patient did not have a tethered spinal cord, and during his surgery, it did not appear that the pre-sacral mass was obstructing the colon, nor did he have severe anal stenosis. Taken together, the previous leads us to believe that our patient's

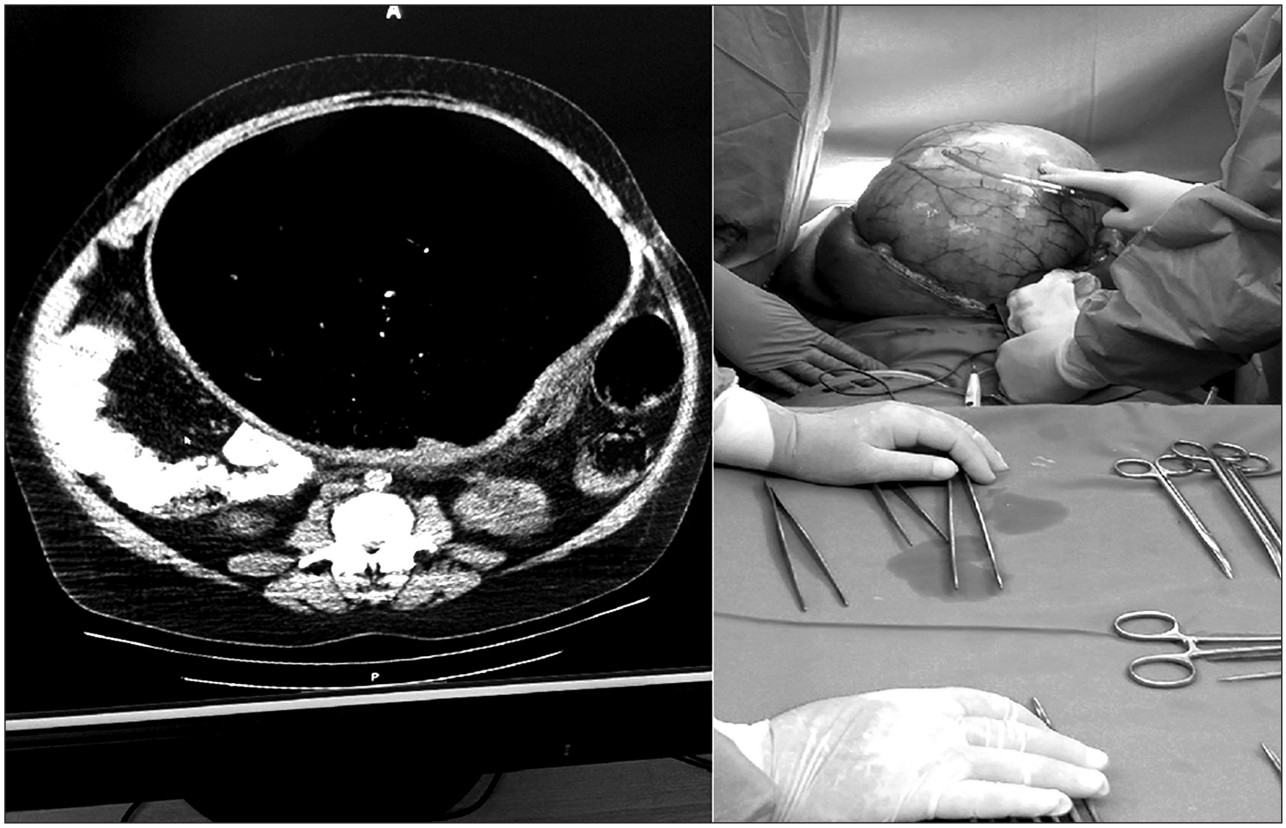

Figure 1. A) Computed tomography scan showing a 25 cm distension of the sigmoid; B) a view of the distended bowel during explorative laparotomy.

condition arose from his history of chronic constipation in combination with a focal neurologic deficit, both of which led to paralytic ileus and extensive distension of the colon. However, this hypothesis would be difficult to verify.

The presence of symptoms and signs of a true bowel obstruction, the lack of any mechanical component causing such obstruction, and the patient's history of systemic sclerosis (SSc), offers a possible explanation for the observed distension of the sigmoid. Chronic intestinal pseudo-obstruction is a rare entity affecting patients with SSc (7). Dysmotility, the key characteristic of SSc in the gastrointestinal tract either due to autonomic dysregulation or due to wall fibrosis (8), along with atony dilation and delayed transit within the bowel, can contribute to impaired gastrointestinal propulsion, further leading to bowel obstruction without any mechanical occlusion in the gut's lumen (7). Large bowel involvement in SSc is rather rare (affecting 20-55% of patients) (7,8), and data concerning its course and management are scarce (9), however we felt that our patient's reduced rectal tone and mobility due to the both the SSc and the CS could have aggravated bowel function to such a degree that the observed sigmoid distension occurred.

The systemic sepsis of our patient could have been caused by bacterial translocation and gut-derived sepsis (10), as no evidence of perforation was observed. Battling sepsis requires lightning-fast action, in terms of the critical need to immediately recognize clinical deterioration and implement treatment. A very balanced decision was made quite early to proceed with surgery, thereby reducing the time from admission to the OR (a total of 4 hours, including the abdominal CT evaluation). All in all, the main goals of early resuscitation, early antibiotic administration, and early source control management were met with success.

Intraoperatively, our patient's cardiac rhythm ranged from sinus tachycardia to atrial flutter and fibrillation.Managing an elevated heart rate accompanied by low blood pressure can be challenging during sepsis . Our patient was known to have AF, which was being treated with anticoagulants and b- blockers, this is why a rate -control strategy that featured a continuous infusion titrated to an appropriate hemodynamic point to avoid adverse effects was preferred.

The drug that we selected for rate control (intra- and postoperatively) for our patient, was Landiolol. It is an ultrashort cardio-selective b1 -blocker. Several trials have been published proving its efficacy and safety in provoking a more potent negative chronotropic action than other b1-blockers do, with less effect on blood pressure (11,12). Although most of the studies refer to the handling of AF in patients with cardiac dysfunction (13,14), some do show improvement of the heart rate with hemodynamic stabilization and no blood pressure reduction in patients with sepsis (15,16). In our patient we used vasopressors for blood pressure control and continuous Landiolol infusion to control ventricular response, doing so without compromising his hemodynamic stability. Regarding heart rate, the target of less than 110 bpm was achieved, and there was no change in MAP. After the removal of the source of infection, the Landiolol infusion (without any added vasopressors) was used at very low dose for rate control in the ICU.

Conclusion

We report this case to increase awareness about CS, to emphasize the importance of considering the syndrome when approaching cases of long-standing refractory constipation, and to highlight the growing role of Landiolol in handling complex septic states.

Resumen

El síndrome de Currarino (CS) estáa caracterizado por una tríada de malformaciones: un defecto sacrococcígeo, una masa en el espacio presacro y de malformaciones del ano o el recto (o ambos). Presentamos el caso de un paciente adulto con CS en shock séptico que se sometió en laparotomía de emergencia a causa de una grave distensión abdominal. Queremos subrayar por un lado la importancia de cirugía inmediata y por otro lado el considerable papel de Landiolol en controlar el ritmo cardiaco del paciente sin empeorar su presión arterial durante este episodio de sepsis y fibrilación auricular.

References

1. Colapinto MN, Vowinckel EA, Colapinto ND. Complete Currarino syndrome in an adult, presenting as a fecalith obstruction: report of a case. Can J Surg. 2003;46(4):303-306.
2. Djordjevic I, Jovanovic D, Grujic B. Currarino Syndroma, initially misdiagnosed and treated like megacolon congenitum. Open Med. 2011 Jan 1;6(6):723-726. https://doi.org/10.2478/s11536-011-0091-4
3. Wald A. Slow Transit Constipation. Curr Treat Options Gastroenterol. 2002;5(4):279-283. doi:10.1007/s11938-002-0050-x
4. Mayr A, Ritsch N, Knotzer H, et al. Effectiveness of direct-current cardioversion for treatment of supraventricular tachyarrhythmias, in particular atrial fibrillation, in surgical intensive care patients. Crit Care Med. 2003;31(2):401-405. doi:10.1097/01.CCM.0000048627.39686.79
5. Walkey AJ, Evans SR, Winter MR, Benjamin EJ. Practice Patterns and Outcomes of Treatments for Atrial Fibrillation During Sepsis: A Propensity-Matched Cohort Study. Chest. 2016;149(1):74-83. doi:10.1378/chest.15-0959
6. Aydoseli A, Akcakaya MO, Aras Y, Dolas I, Yanar H, Sencer A. Anterior sacral meningocele in a patient with currarino syndrome as a cause of ileus. Br J Neurosurg. 2013;27(6):833-835. doi:10.3109/02688697.2013.785476
7. McFarlane IM, Bhamra MS, Kreps A, et al. Gastrointestinal Manifestations of Systemic Sclerosis. Rheumatology (Sunnyvale). 2018;8(1):235. doi:10.4172/2161-1149.1000235
8. Nagaraja V, McMahan ZH, Getzug T, Khanna D. Management of gastrointestinal involvement in scleroderma. Curr Treatm Opt Rheumatol. 2015;1(1):82-105. doi:10.1007/s40674-014-0005-0
9. Mecoli C, Purohit S, Sandorfi N, Derk CT. Mortality, recurrence, and hospital course of patients with systemic sclerosis-related acute intestinal pseudo-obstruction. J Rheumatol. 2014;41(10):2049-2054. doi:10.3899/jrheum.131547
10. Deitch EA, Xu D-Z, Lu Q. Gut lymph hypothesis of early shock and trauma-induced multiple organ dysfunction syndrome: A new look at gut origin sepsis. J Organ Dysfunct. 2006;2(2):70-79. doi:10.1080/17471060600551772

11. Atarashi H, Kuruma A, Yashima M, et al. Pharmacokinetics of landiolol hydrochloride, a new ultra-short-acting beta-blocker, in patients with cardiac arrhythmias. Clin Pharmacol Ther. 2000;68(2):143-150. doi:10.1067/mcp.2000.108733
12. Sasao J, Tarver SD, Kindscher JD, Taneyama C, Benson KT, Goto H. In rabbits, landiolol, a new ultra-short-acting beta-blocker, exerts a more potent negative chronotropic effect and less effect on blood pressure than esmolol. Can J Anaesth. 2001;48(10):985-989. doi:10.1007/BF03016588
13. Yamashita T, Nakasu Y, Mizutani H, Sumitani K. A prospective observational survey on landiolol in atrial fibrillation/atrial flutter patients with chronic heart failure - AF-CHF landiolol survey. J Cardiol. 2019;74(5):418-425. doi:10.1016/j.jjcc.2019.05.012
14. von Haehling S, Bělohlávek J, Er F, Gassanov N, Guarracino F, Bouvet O. Landiolol for rate control management of atrial fibrillation in patients with cardiac dysfunction. Eur Heart J Suppl. 2018;20(Suppl A):A19-A24. doi:10.1093/eurheartj/sux035
15. Okajima M, Takamura M, Taniguchi T. Landiolol, an ultra-short-acting β1-blocker, is useful for managing supraventricular tachyarrhythmias in sepsis. World J Crit Care Med. 2015;4(3):251-257. Published 2015 Aug 4. doi:10.5492/wjccm.v4.i3.251
16. Arita Y, Segawa T, Yamamoto S, Hasegawa S. Landiolol is effective for the treatment of tachycardia-induced cardiogenic shock in patients during septic shock therapy. BMJ Case Rep. 2017;2017:bcr2017222268. Published 2017 Nov 1. doi:10.1136/bcr-2017-222268