

Peer review file

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Reviewer A

Comment: There are some typos and grammatical errors in your article. Please ask an expert who is proficient in medical English to review and improve your article.

Reply: We thank the reviewer for this comment. The article was proofread by two separate individuals with knowledge in medical English. All changes to the article, including spelling and grammatical errors, are in red font.

Reviewer B

Comment 1: In the abstract and conclusion, the word excellent cosmetic result etc. is mentioned.

Is that the opinion of the patient or the surgeon, since the patient is left with a rather large scar (see figures)?

Reply 1: We thank the reviewer for this comment. This subjective statement was removed from our conclusions in the mentioned locations. It does still remain in those as cited in outside sources.

Changes in the text: we have modified our text as advised (see page 9, line 225-227): In conclusion, **our retrospective results suggest that the surgical correction of CS syndrome be pursued at this time only with the goal of cosmetic improvement, and although patients reported subjective improvement in dyspnea related symptoms, significant objective clinical benefit was not appreciated in the patients studied.**

Comment 2: line 62 is not clear. If I get it right, one person had for cosmetic reasons in the past a Nuss bar implant. To state that she refused surgery for cosmetic reasons is confusing and in that case the numbers described don't add up.

Reply 2: We thank the reviewer for this comment. This phrase was made clearer, with

one of the two patients not undergoing surgical correction having undergone prior Nuss procedure with no benefit. This patient was not one of the two surgical candidates, but information regarding her course was included as part of the retrospective review.

Changes in the text: we have modified our text as advised (see page 4, line 81-83): Two of the four patients were asymptomatic and refused surgical correction **for what they viewed as purely** cosmetic purposes (Fig. 2). One of the two **was misdiagnosed with PE type of deformity and corrected** with Nuss procedure three years prior with no significant clinical benefit.

Comment 3: line 67. Also, case series can be subject to necessary medical ethical approval. What you probably mean is exception on base of retrospective provider health care evaluation.

Reply 3: We thank the reviewer for this comment. Yes, case series was incorrectly used here and replaced with a retrospective study.

Changes in the text: we have modified our text as advised (see page 4, line 85-88): **As this was a retrospective review, approval from a local ethics committee was not required. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from all patients.**

Comment 4: Have a look at the used frase 10 000. Space less...

Reply 4: We thank the reviewer for this comment. This formatting error was corrected.

Changes in the text: we have modified our text as advised (see page 3, line 51): PC is seen less frequently than PE, comprising about 5–15% of patients with pectus abnormalities and occurs in about **1:1000–1:10,000** of all live births with a male predominance (4:1)^{1,2}.

Comment 5: line 70. I missed the time period used for the literature search.

Reply 5: We thank the reviewer for this comment. The time period was inserted into the literature review section.

Changes in the text: we have modified our text as advised (see page 4, line 93-94): A total of 22 appropriate full-text case reports and case series in English language were identified through database and reference searches from 1952 to 2019 (Table 2).

Comment 6: line 186. What happened to the dyspnea in patient 1 and the exertional dyspnea in patient 2 after the surgery. Did it improve and if so, is the statement in your manuscript that surgery is only for cosmetic reasons than correct or partially correct? I would add that to the discussion.

Reply 6: We thank the reviewer for this comment. Discussion was created regarding the use of the procedure solely for cosmesis with noted patient subjective improvement in symptoms as noted. Follow up with objective data regarding clinical symptom resolution was not completed.

Changes in the text: we have added data regarding symptoms in postoperative period (see page 5, line 116-117 and page 5 line 127) and changed conclusion as advised (see page 9, line 225-227): The follow-up period was 23 months, with subjective resolution of her symptoms and no recurrence of the deformity noted.

The follow-up period was 34 months, with subjective resolution of her exertional dyspnea and no recurrence of the deformity noted (Fig. 7C-D).

In conclusion, our retrospective results suggest that the surgical correction of CS syndrome be pursued at this time only with the goal of cosmetic improvement, and although patients reported subjective improvement in dyspnea related symptoms, significant objective clinical benefit was not appreciated in the patients studied.

Comment 7: Doe the figure 1 have copyright and if so does it need a mention of the owner?

Reply 7: We thank the reviewer for this comment. Figure 1 is an original image with no copyright associated.

Comment 8: A suggestion would be to limited the number of figures for example Fig1 - Fig 7 AB - Fig 2 - Fig 6 BC - Fig 7 CD. It would also make it a more logic picture story enhancing your manuscript.

Reply 8: We thank the reviewer for this comment. We removed Fig. 3 and 4. Other figures were streamlined to allow for a clearer presentation and flow.

Reviewer C

Comment 1: Title: Please include the core message of the report. The core message beside literature review are diagnosis and treatment of CSS.

Reply 1: We thank the reviewer for this comment. Title was adjusted as suggested.

Changes in the text: we have modified our title as advised (see page 1, line 1):
Currarino-Silverman Syndrome: **Diagnosis and Treatment of** Rare Chest Wall Deformity.

Comment 2: Key words: Core message (treatment and diagnosis of CSS) not included

Reply 2: We thank the reviewer for this comment. Key words were added as suggested.

Changes in the text: we have modified our title as advised (see page 2, line 41):
diagnosis and treatment of Currarino-Silverman syndrome, pouter pigeon chest, chondromanubrial deformity, pectus arcuatum, case series.

Comment 3: Introduction: the aim of the present paper remains diagnosis and treatment of CSS. So, the first paragraph does not provide essential insights and is not really relevant for the topic. The paragraph about the classification is ok, but what is the difference between pectus arcuatum and CSS? Is there a difference? I would suggest some thoughts also about the treatment options in anterior chest wall deformities, especially regarding the conservative treatment (vacuum bell and FMF compressive orthosis). Why they do not work in a s-shaped sternum?

Reply 3: We thank the reviewer for this comment. Elaboration was completed regarding failure of conservative therapy in deformity related to CS, as well as how CS deformities compare and relate to other pectus deformities. There is no difference between pectus arcuatum and CSS. However, CSS can be easily mistaken with PE since both deformities appear in almost similar fashion, but surgical approaches are very different.

Changes in the text: We have modified our text as advised (see page 9, line 218-220): In comparison to PE, a number of less invasive techniques have been developed for the correction of PC, including the Abramson procedure and its modifications, along with nonsurgical options such as observation, orthotic bracing, and dynamic compression^{2,31}. However, due to the extreme rarity of the disease, challenging deformity and variable anatomy of fused sternum, there are no clear guidelines in the treatment approaches. **This unique growth pattern contributes to almost universal failure of conservative treatment options like the vacuum bell or a compressive orthosis.**

Comment 4: Patients/Methods:

Please provide information about written consent from the patient for picture publication. Is written consent available for review?

Reply 4: We thank the reviewer for this comment. Written consent was obtained for all patients regarding all published materials.

Changes in the text: We have added this information (see page 4, line 84-88): **As this was a retrospective review, approval from a local ethics committee was not required. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from all patients.**

Comment 5: - Patient 1: Please provide type/mark of used steel wires

Reply 5: We thank the reviewer for this comment. The specific type of wires with citation was added.

Changes in the text: We have modified our text as advised (see page 5, line 113): Osteosynthesis of the lower sternum osteotomy site was then accomplished using two parallel **316L stainless steel surgical monofilament wires (Ethicon, USA) (Fig. 5).**

Comment 6: - Is a postoperative evaluation of quality of life or satisfaction questionnaire available?

Reply 6: We thank the reviewer for this comment. No postoperative questionnaire regarding quality of life was administered to patients.

Comment 7: - I would prefer a more precise description of the surgical technique used to correct the two female patients!

Reply 7: We thank the reviewer for this comment. More precise description of surgical procedure was formed for both patients.

Changes in the text: we have modified our text as advised (see page 5, line 104-114): Surgical correction was performed through a transverse submammary skin incision **in respect to the lines of Langer. Musculocutaneous flaps were dissected bilaterally up to the level of the sternal notch with detachment of the pectoralis musculature and retracted laterally.** Subperichondrial resections of the 2nd–5th costal cartilages **were completed bilaterally with preservation of perichondrium to avoid chest wall weakness postoperatively. Careful attention was also given to the growth centers of the sterno-chondral and costo-chondral junctions to avoid development of a restrictive deformity.** In the projection of maximal protrusion of the sternal body, at the level of the third rib and second intercostal space, a two-level wedge-shaped transverse osteotomy was subsequently performed using an oscillating saw preserving the integrity of the posterior lamina. Remodeling of the sternum was performed by applying caudal pressure at the top of the deformity and anterior traction at the sternal body. Osteosynthesis of the lower sternum osteotomy site was then accomplished using two parallel **316L stainless steel surgical monofilament wires (Ethicon, USA) (Fig. 5).** **The edges of the divided pectoralis musculature were approximated with interrupted suture followed by closure of the skin flaps.**

Comment 8: - Has cardiac compression after correction of the lower part of the sternum been evaluated preoperatively?

Reply 8: We thank the reviewer for this comment. Clinical outcomes with objective data were not available postoperatively to compare with preoperative findings.

Comment 9: Literature review:

- Have studies been excluded from the review?

Reply 9: We thank the reviewer for this comment. No specific studies were excluded from this literature review.

Comment 10: - In general, those are very small series with short follow-up; please comment on the outcome/validity?

Reply 10: We thank the reviewer for this comment. Comments were added regarding the short-term outcome of the literature available, and likely situations related to this.

Changes in the text: we have modified our text as advised (see page 4, line 94-96):

Much of the identified literature is limited by relatively short term follow up, likely secondary to the younger patient population and the lack of symptoms both pre- and post-operatively.

Comment 11: Discussion:

- Line 131-133: Please provide information about acquired thoracic dystrophy if cartilage resection is performed in a too extensive manner or in a too young age

Reply 11: We thank the reviewer for this comment. The complications associated with this were added to the discussion.

Changes in the text: We have modified our text as advised (see page 5, line 106-108 and page 7, line 164-165): Subperichondrial resections of the 2nd–5th costal

cartilages were completed bilaterally with preservation of perichondrium to avoid chest wall weakness postoperatively. Careful attention was also given to the growth centers of the sterno-chondral and costo-chondral junctions to avoid development of a restrictive deformity.

Late puberty or adulthood was indicated as a preferred age for correction since cartilages resection will be performed when rib growth ends^{7,18,23}. It is important to keep in mind the potential for thoracic dystrophy should cartilage resection be carried out at too young an age, or too extensively.

Comment 12: - Line 140: ...with PC deformity or CSS?

Reply 12: We thank the reviewer for this comment. Corrected to CS syndrome.

Changes in the text: We have modified our text as advised (see page 7, line 173):

Thus, cardiovascular evaluation should be performed in all patients with CS syndrome.

Comment 13: - Line 140: ...Chest CT with 3-D reconstruction or MRI? Please

explain why MRI is more indicated in adolescent people than CT-scan

Reply 13: We thank the reviewer for this comment. Reasoning behind each imaging modality was added to the discussion.

Changes in the text: We have modified our text as advised (see page 7, line 173-174): Chest CT with 3-D reconstruction, **or MRI in children given altered tissue density and radiation concern**, is the best preoperative imaging for evaluation of CS patients.

Comment 14: -Please refer to the work of Prof. Jean-Marie WIHLM, world famous thoracic surgeon and in the field of the correction of pectus arcuatum by sterno-costochondroplasty! ANZ J Surg. 2019 Oct;89(10):1342-1344.

Reply 14: We thank the reviewer for this comment. This paper describes the surgical correction of chondrogladiolar type, which is out of the scope of our study. However, we cited a different work of Prof. Wihlm in our paper. Please, see reference number 27.

Comment 15: Conclusion:

- Place limitations statement not in the conclusion

Reply 15: We thank the reviewer for this comment. Limitations moved to discussion from conclusion.

Changes in the text: we have modified our text as advised (see page 9, line 221-224): **This review was of a single institution with limitations inherent of a retrospective study. This review is without a comparison analysis, since only one surgical technique was performed. Case numbers for comparison are difficult to ascertain given the rarity of the disease process. The asymptomatic nature of the deformity also leads to a degree of hesitancy regarding surgical correction, again limiting data.**

Comment 16: - Please provide thinks to be done next and another important message: every single surgeon should not treat CSS. Its correct management requests a dedicated team (ideally thoracic and pediatric surgeon or pectus surgeon) with specific expertise in this field!

Reply 16: Recommendations regarding location for intervention, as well as next steps

in research added to conclusion.

Changes in the text: We have modified our text as advised (see page 9, line 229-233): **Given the rarity of the deformity, it's surgical correction should be completed by a multidisciplinary team, including thoracic reconstructive surgeons with experience in pectus deformities. Future study could include more extensive pre-operative workup and data collection to compare with postoperative values, to ascertain whether a clinical benefit may also accompany the cosmetic result achieved by surgical intervention.**

Comment 17: Picture 1: Not very useful. Provide source of this picture! Provide picture that allow to better understand CSS.

Reply 17: Picture 1 is an original illustration created by a hired medical professional to best represent in simple terms the defect of CS syndrome.