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Diagnostic value of chest ultrasound in children with cystic fibrosis.

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Abstract

Cystic fibrosis (CF) is one of the most common genetic disorders in the Caucasian population. The disease has a progressive course and leads to reduced life quality and life expectancy. Standard diagnostic procedures used in the monitoring of CF patients, include methods exposing patients to the ionizing radiation. With increasing life expectancy in CF the cumulative dose of ionising radiation increases, prompting clinicians' search for safer imaging studies. Despite its safety and availability lung ultrasound (LUS) is not routinely used in the diagnostic evaluation of CF patients.

The aim of the study was to evaluate the diagnostic value of LUS in children with CF compared to chest X-ray, and to assess the diagnostic value of the recently developed LUS score - CF-USS (Cystic Fibrosis Ultrasound Score).

LUS was performed in 48 CF children aged from 5 to 18 years (24 girls and 24 boys). LUS consisted in the assessment of the pleura, lung sliding, A-line and B-line artifacts, "lung rockets", alveolar consolidations, air bronchogram and pleural effusion. Chest radiography was performed in all patients and analyzed according to the modified Chrispin-Norman score. LUS was analyzed according to CF-USS.

Correlation between the CF-USS and the modified Chrispin-Norman scores were moderate (R=0.52, p=0.0002) and strong in control studies . In 75% of patients undergoing LUS, small areas of subpleural consolidations were observed, not visible on X-rays. At the same time, LUS was not sensitive enough to visualize bronchial pathology, which plays an important role in assessing the disease progression.

Conclusions:

LUS constitutes an invaluable tool for the diagnosis of subpleural consolidations. CF-USS results correlate with conventional x-ray modified Chrispin–Norman score. LUS should be considered an accessory radiographic examination in the monitoring of CF patients, and CF-USS may provide clinicians with valuable information concerning the disease progression.

Introduction

Cystic fibrosis (CF) is one of the most common autosomal recessive hereditary life-shortening disorders in Caucasian populations [1,2]. The disease is caused by the mutation of gene coding CFTR protein (*Cystic Fibrosis Transmembrane Conductance Regulator*), leading to the production of dense mucus in the airways and exocrine glands and the impairment of their function. The main affected systems comprise respiratory and digestive systems, and the chronic pulmonary disease remains the main cause of morbidity and one of the most important prognostic factors in CF [1,3,6]. Chronic inflammation due to impaired mucocilliary clearance and mucus impaction in the airways results in bronchiectasis and progressive lung tissue destruction [5].

Lung evaluation in CF patients traditionally avails of imaging studies and among these the most commonly used remains chest x-ray. Early in the course of disease the radiologic picture might reveal no abnormalities. Along with the disease progression lung hyperinflation and increased bronchial markings appear, followed by chest infiltrates, atelectasis and bronchiectasis [1,6].

The need for objective tools for the evaluation of patients has prompted the development of x-ray scoring systems including Brasfield score [7] Northern score [4] Chrispin-Norman score [8] and its modified version [9]. These scoring systems are used for the monitoring of disease progression, evaluation of different therapies as well comparison of patients' outcomes between the treatment centres [4,8–14].

The most accurate radiographic diagnostic modality in CF, so called „golden standard" that allows for qualitative and quantitative evaluation of lung involvement, even very early in the course of the disease remains computed tomography (CT) [6]. CT due to its high resolution allows visualisation of the detailes that are not visible in the plain chest x-ray [10]. In CF patients CT enables visualisation of bronchial wall and peribronchial thickening, intralobular nodules, bronchiolitis, so called „tree in bud" sign, air trapping, bronchiectasis, mucus impaction, microabscesses, infiltrates, atelectasis, enlarged lymph nodes and widening of pulmonary artery with narrowing of peripheral vessels [5,15,16]. The role of CT in CF patients was confirmed in studies reporting on correlation of CT scans with patients outcomes [17]. For quantitative, objective evaluation of CT results in CF patients scoring systems were also developed with the most popular Bhalla score [18].

Disadvantage of CT scanning is a relatively high dose of ionising radiation. The risks of cancer related to lifetime exposure to radiation made clinicians look for imaging modalities with the lowest or ideally no radiation [19]. Ultrasound (US) is currently one of the most important and

88   most frequently used imaging techniques [20]. Considering this, lung ultrasound (LUS) as a safe,
89   non-invasive, widely available and cheap technique might constitute an important tool in the
90   diagnostic protocols of children with CF [21]. Despite this fact there are few existing reports on
91   LUS application in CF patients. There are only two reports published as abstracts by Ciuca et al on
92   LUS in CF as compared to CT scans [22,23].

93   LUS examination comprise evaluation of pleural line and lung sliding [24–28], analysis of
94   the artefacts that are present in normal lung, like „the bat sign" [24,25,29,30] and the A-line
95   artefacts [25,28,31,32] as well as in pathological conditions (the B-line, Z-line and I-line artefacts)
96   and evaluation of thoracic wall structures. The B-lines are vertical, well defined hyperechogenic
97   lines, arising from the pleural line, spreading out without fading to the edge of the screen, similar to
98   laser beam or „comet tail" artefact [25,33,34]. Multiple B-lines are typical for interstitial lung
99   disease [35–37]. Seen together they are described as „lung rockets" artefact [28,32,37]. Multiple
100   coalescent B-lines in the absence of A-lines with visible lung sliding constitute so called „white
101   lung" image [37–39]. Alveolar consolidation can be diagnosed with LUS provided their peripheral
102   localisation and according to the literature reports that is the case in up to 98.5% of cases [24,29,32].

Objective

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105   The aim of our report was to evaluate the diagnostic value of chest ultrasound in children with CF
106   as compared to plain x-ray, as well as to assess the diagnostic value of the recently developed LUS
107   score - CF-USS (Cystic Fibrosis Ultrasound Score).

Material

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110   We enrolled 48 Caucasian patients (24 males) aged 5 to 18 years diagnosed with CF who
111   were admitted to the Pulmonology Department for scheduled annual diagnostic workup. Patients
112   underwent chest ultrasound and plain x-ray, and the time interval between the studies were not
113   longer than 72 hours.
114   In all the studied children CF was confirmed by two positive sweat test results ant genetic studies
115   (two pathogenic mutations). All the patients and their parents gave informed consent for the study.
116   Exclusion criteria comprised severe immunosuppression, lack of consent and time interval longer
117   than 72 hours between the studies. The study design was accepted by the Bioethical Committee of
118   Poznań University of Medical Sciences. In Table 1 we presented the characteristic of the studied
119   patients.

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121   **Table 1.** Patients characteristic

Number of patients	48	
Males/females	24 / 24	
	Mean ± SD	Median (minimum-maximum)
Age (years)	11.9 ± 3.9	12 (5-18)
BMI	16.7 ± 2.9	15.7 (10.2-24.2)
FEV_1 (L)	1.9 ± 0.8	1.7 (0.4 - 4.3)
BMI <3c N(%)	7 (14.6)	
Pancreatic sufficient N (%)	9 (18.8)	
F508del homozygous N(%)	21 (43.8)	
F508del heterozygous N(%)	18 (37.5)	
Chronically infected with *P. aeruginosa*	21 (43.8)	
$FEV_1 \geq 80\%$ normal value	32 (66.7)	
$FEV_1 \leq 40\%$ normal value	1 (2.1)	

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Methods

Radiographic imaging

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127   X-rays were performed with an analogue apparatus Axiom Iconos R 100 (Siemens
128   Healthcare), in posteroanterior projection during suspended inspiration. Technical parameters of the
129   images (including use of grid, the source image receptor distance, dose of radiation) were
130   individually adjusted for every studied patient in concordance with ALARA (As Low As
131   Reasonably Achievable) principle in order to achieve best possible images using the lowest
132   radiation dose. No lateral x-rays were performed. X-rays were independently evaluated by two

133   board certified paediatric radiologists with experience in CF. Chest x-rays were evaluated using

134   modified Chrispin-Norman score [9].

Chest ultrasound (LUS)

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137   Chest ultrasound was performed with iU22 apparatus (Philips, Biothel United States) using

138   linear probe of 5 – 12 mHz (L12-5) frequency and depending on the patients' age with either

139   convex probe of 1 - 5 mHz (C5-1) frequency, convex probe of 4 - 9 mHz (C9-4) frequency or

140   microconvex probe of 5 – 8 mHz (C8-5) frequency through longitudinal and transverse sections of

141   anterior, lateral and posterior wall of the chest. Preliminary preset was soft tissue excluding artefact

142   reduction options (SonoCT, XRes). Doppler imaging was used for the evaluation of vascularisation

143   of the inflammatory changes.

144   Patients were examined in the sitting position. The studies were performed by two board certified

145   paediatric radiologists with experience in LUS and CF.

146   In every patient we evaluated the quality (free flowing or organised, localization) and quantity

147   (fluid layer in millimetres) of any fluid present in the pleural space, the shape and thickness of the

148   pleural line, the lung sliding sign, A-lines and B-lines artefacts (their number, localisation and

149   morphology, including single ones as well as „lung rockets" complexes and "white lung" images)

150   and alveolar consolidations (their number, dimensions, localisation, morphology, presence of

151   bronchogram and its characteristic (air or fluid) and vascularisation).

152   LUS results were classified according scoring system developed by the authors: CF-USS

153   *(Cystic Fibrosis Ultrasound Score)* devised on the basis of modified Chrispin-Norman score and

154   bronchiolitis score reported by Caiulo and collaborators [39,40]. Scores are calculated separately for

155   anterior and posterior surface of right and left half of the thorax. Each part can be scored from 0 to 2

156   points for irregularities of pleural line, single and complex B-line artefacts, alveolar consolidations

157   and the presence of fluid in the pleural space with the maximum score of ten for each part and 40 in

158   total. The higher the score, the more advanced the disease process (Table 2).

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165   **Table 2.** Cystic Fibrosis Ultrasound Score

Characteristic	Intensity		
Pleural irregularities	Absent	Present	Present+ pleural thickening
Right lung: anterior surface	0	1	2
posterior surface	0	1	2
Left lung: anterior surface	0	1	2
posterior surface	0	1	2
Focal B-line artefacts	Absent / few (≤6)	Some (7-14)	Many (≥15)
Right lung: anterior surface	0	1	2
posterior surface	0	1	2
Left lung: anterior surface	0	1	2
posterior surface	0	1	2
Coalescent B-line artefacts	Absent	Fused	„Lung rockets"
Right lung: anterior surface	0	1	2
posterior surface	0	1	2
Left lung: anterior surface	0	1	2
posterior surface	0	1	2
Subpleural consolidations	Absent (≤6)	Some (7-14)	Multiple or extensive (≥15)
Right lung: anterior surface	0	1	2
posterior surface	0	1	2
Left lung: anterior surface	0	1	2
posterior surface	0	1	2
Pleural fluid	Absent	Obliterating phreno – costal angle	In phreno – costal angle and along the chest wall
Right lung: anterior surface	0	1	2
posterior surface	0	1	2
Left lung: anterior surface	0	1	2
posterior surface	0	1	2

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167 ## Statistical analysis

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169 Statistical analysis was performed with Statistica software (version 12; StatSoft). Data
170 distribution was evaluated with Shapiro-Wilk test. For data with normal distribution we used t
171 Student test for paired and independent variables. For data that do not meet the normal distribution
172 assumptions Spearman's rank correlation coefficient was calculated. P value of <0.05 were
173 considered statistically significant.

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175 # Results

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177 ## Comparison of LUS and chest x-ray images

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179 Pulmonary disease was evaluated radiologically with modified Chrispin – Norman score for
180 x-rays and CF-USS score. The patients' results are shown in Table 3, Figures 1 and 2. Statistical
181 analysis has shown positive correlation between the two scoring systems
182 (R Spearman= 0.52, p=0.0002) (Fig 3).

183

184 **Table 3.** Comparison of two scoring systems

Parametr	Modified Chrispin – Norman score	CF-USS
Number of patients	48	48
Mean ± SD	8.7 ± 8.0	4.4 ± 4.1
Median (minimum-maximum)	7 (0-28)	4 (0-16)

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187 **Fig 1. The modified Chrispin-Norman scores**.

188 **Fig 2. The CF-USS scores.**

189 **Fig 3. Correlation between CF-USS and the modified Chrispin-Norman scores.**

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191 Fine subpleural consolidation were seen in LUS in 36 patients (75%). Abnormalities seen in
192 LUS in the studied patients classified according to CF-USS are presented in Table 4.

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194 **Table 4.** Number of patients in the studied group with abnormalities according to the CF-USS score.

Characteristic	Number of patients	Percentage (%)
Pleural irregularities	1	2
Focal B-line artefacts		
Some	33	69
Many	4	8
Fused B-line artefacts		
Coalescent	19	40
„Lung rockets"	1	2
Subpleural consolidations		
Few	28	58
Multiple or extensive	8	17
Pleural fluid		
In costo – phrenical angle	9	19
And along the chest wall	2	4

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198 In nine patients LUS and x-ray were performed twice on two different occasions. There were no

199 statistically significant differences between the x-rays in modified Chrispin – Norman score

200 (15.22±2.71 vs. 10.78±3.00; p=0.06) and LUS in CF-USS score (7.56±1.58 vs. 5.33±1.86; p=0.29)

201 (Fig 4 and Fig 5). Statistical analysis for the repeated LUS and x-ray examinations showed positive

202 correlation for the two studies (R Spearman= 0.81, p=0.01) (Fig 6).

203 **Fig 4. Modified Chrispin – Norman scores for the repeated studies.**

204 1. Modified Chrispin – Norman scores in the first study; 2. Modified Chrispin – Norman

205 scores in the repeated study

206 **Fig 5. CF-USS scores for the repeated studies.**

207 1. CF-USS scores in the first study; 2. CF-USS scores in the repeated study

208 **Fig 6. Correlation between the CF-USS score and the modified Chrispin-Norman score in the**

209 **repeated studies.**

210 In the figures 7-11chest x-ray and LUS images of the studied patients are presented.

211

212 **Fig 7.**

213 Mediastinal shift, atelectasis and pleural effusion on the right, linear and cystic opacities,

214 bronchiectasis, consolidations. Hyperinflation of the left lung, with nodular and linear interstitial

215 opacities. 27 points in modified Chrispin-Norman score.

216 **Fig 8.**

217 LUS image of the patient from Fig 7, linear probe. Regions of consolidation and atelectasis. 17

218 points in CF-USS.

219 **Fig 9.**

220 Linear opacities and regions of consolidations in the middle right and lower left field. Fine

221 peribronchial infiltrates in the lower right and middle left field. Hyperinflation of both lungs. 15

222 points in modified Chrispin-Norman score.

223 **Fig 10.**

224 LUS image of the patient from Fig 7, linear probe. Large area of consolidation in the right upper

225 lobe. 5 points in CF-USS.

226 **Fig 11.**

227 LUS image of the patient from Fig 7, linear probe. Fine subpleural consolidation.

228 # Discussion

229

230   Cystic fibrosis is a life-shortening genetic disorder, involving respiratory system and

231 requiring chronic therapy. In the course of the disease patients suffer recurrent exacerbations, that

232 affect patients quality of life and survival. Radiology plays a significant role in patients' follow-up,

233 enabling monitoring of the disease, response to treatment as well as the diagnosis of exacerbations

234 [41]. Unfortunately conventional x-rays, especially numerous, repeated in the course of disease add

235 up to cumulative ionising radiation dose. Diminishing radiation exposure, by looking for alternative

236 diagnostic modalities, should be considered as one of the goals of contemporary medicine.

237   Chest ultrasound has not been routinely used in the monitoring of lung disease in paediatric

238 patients with CF, despite its lack of radiation, availability and safety. Considering this we wanted to

239 compare diagnostic value of LUS with conventional chest x-rays assessed according to the modified

240 Chrispin – Norman score. There are no other studies comparing modified Chrispin – Norman score

241 with chest ultrasound in CF paediatric patients. Furthermore, we developed our own ultrasound

242 scoring system: CF-USS to make the comparison more feasible

243   For the evaluation of chest x-ray we chose modified Chrispin – Norman score as it uses only

244 antero – posterior projections for the evaluation of the hyperinflation of the chest based on the shape

245 of the thorax, diaphragm location and lung hyperlucency resulting from air-trapping [9]. That stays
246 in agreement with the Benden et al. data and allows for the diminished radiation dose while
247 avoiding the lateral projection [9]. Terheggen-Lagro and colleagues in their study compared six
248 different clinical and radiological scoring systems (Schwachman – Kulczycki score, Chrispin-
249 Norman score, modified Chrispin-Norman score, Brasfield score, Wisconsin score and Northern
250 score) and demonstrated their clinical utility in different clinical settings [14]. Authors proved, that
251 radiographic scoring systems in the CF patients, especially modified Chrispin-Norman score are
252 characterized by low interobserver variability and correlate with pulmonary function tests results as
253 well as clinical features.

254   The aim of the study was to compare the results of x-ray scoring system with chest
255 ultrasound scoring system. Furthermore we constructed a novel chest ultrasound score for the
256 evaluation of CF paediatric patients (CF-USS). The score has been developed based on the
257 experience of Caiulo and colleagues, who used LUS in patients with bronchiolitis, the pathology,
258 that among others is also present in the CF patients [5,15,40,42].

259   LUS was performed at the same time as the chest x-ray. In nine patients LUS were
260 performed twice on two different occasions. The most commonly seen pathological features were
261 B-line artefacts of different number and intensity. B-line artefacts might be seen in a normal lung
262 and are not considered pathological as long as their number does not exceed 2 in a single transverse
263 scan with a convex probe and 6 in a single longitudinal scan with a high resolution linear probe
264 [32].

265   Clinical relevance of B-line artefacts is quite wide and has recently been covered in an
266 excellent review by Dietrich and colleagues [43]. The authors believe, that B-line artefacts can be
267 caused by multiple factors, and be present in lung oedema, heart failure, lung interstitial diseases,
268 infections, acute respiratory distress syndrome (ARDS) or lung injury. B-line artefacts are the sign
269 of increased lung density due to the loss of the lung tissue aeration. Chiesa and colleagues found B-
270 line artefacts in 37% of elderly studied as compared to 10% of healthy young adults [44]. Correct
271 B-line artefacts interpretation should account for the evaluation of other LUS signs and clinical data.
272 In the pathological conditions B-line artefacts may be useful for the monitoring of treatment. The
273 influence of technical factors on the appearance of B-line artefacts still remains to be elucidated
274 [43].

275   In our LUS scoring system (CF-USS) B-line artefacts are divided into focal (few, some and
276 many) and coalescent (absent, fused and "lung rockets"). The scoring system reflects intensity and
277 variability of the lung pathology, known as B-line artefacts. Despite statistically significant
278 correlation between the two studied scores in our material, we believe that true clinical significance

279   of B-line artefacts, in a single LUS examination without clinical data has important limitations. In

280   the children with CF B-line artefacts should be evaluated in the context of disease progression,

281   documenting their numbers and localisation [45].

282   Another pathology seen in LUS are subpleural consolidations. Very fine 3 to 4 mm in

283   diameter subpleural consolidations may be present in healthy children in the first few years of life

284   [20,46]. In children with CF however they have important clinical implications. Dense mucus is

285   blocking the airways, including bronchiole, leading to focal atelectasis and hyperinflation, and

286   resulting in recurrent infections. Peripheral mucus plugs are causing small foci of inflammation,

287   that might progress into disease exacerbations [10]. There are several studies illustrating the fact,

288   that structural changes seen in radiologic examinations may be seen ahead of pulmonary function

289   deterioration [47]. In CF-USS subpleural consolidations were classified as: absent, few and multiple

290   or extensive. In 75% of the studied patients subpleural consolidations were seen in LUS and in 17%

291   the changes were multiple or wider than 10 mm. None of the changes smaller than 10 millimetres

292   were seen in conventional radiograms. In our opinion subpleural consolidations, similarly to B-line

293   artefacts cannot be evaluated without clinical data. Brody and colleagues reported, that in stable CF

294   patients subpleural consolidations should be monitored, as they may lead to clinical deterioration

295   and decline in pulmonary function test results [47]. In patients with bronchopulmonary disease

296   exacerbations diagnosing and monitoring of subpleural consolidations with LUS may limit the

297   number of x-rays performed and in consequence radiation exposure.

298   CF-USS also comprised the evaluation of pleural fluid which seems reasonable to perform

299   in patients suspected of pleural complications regardless of conventional x-ray results. In the

300   studied group in the 23% of patients we have documented the presence of fluid in the pleural space,

301   majority of them having just the small amount in the costo – phrenical angle. Pleural irregularities

302   were seen in only one of the studied patients. Caiulo and colleagues reported in their bronchiolitis

303   study pleural irregularities in 25% of patients that have been disappearing in the course of follow up

304   [40]. The reason for the disparity of our results might be the fact, that bronchiolitis is not present an

305   all patients with cystic fibrosis.

306   In 9 patients we repeated LUS and conventional x-rays in stable condition in the course of 2

307   years follow up. There were no statistically significant differences between either LUS or x-rays.

308   Spearman rank correlation coefficient between x-ray and LUS was higher in the second series of

309   studies.

310   We believe that LUS is an important diagnostic tool, accessory to conventional x-rays

311   enabling monitoring of the disease process. We do acknowledge however the limitations of CF-USS

312   scoring system. Our material is small and the study was conducted in a single CF centre – we do

313 hope however, that the study will be continued in the future. The most important limitation remains
314 inability of visualisation of consolidations separated from pleura as well as of airway pathologies
315 (bronchiectasis, mucus plugs) that constitute the mainstay of lung pathology in CF pulmonary
316 disease. Nevertheless, due to its safety, non-invasiveness and availability we hope that CF-USS will
317 find its place in long term monitoring of the disease, response to treatment and the risk of
318 exacerbations
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Conclusions

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322   1. LUS should be an accessory radiographic examination in the scheduled follow-up visits in
323   cystic fibrosis paediatric patients, and CF-USS scoring system may provide clinicians with
324   valuable informations concerning the disease progression and exacerbations' risk.
325   2. CF-USS results correlate with conventional x-ray modified Chrispin – Norman score.
326   3. LUS constitute an invaluable tool for the diagnosis of subpleural consolidations
327   4. LUS limitations remain inability to visualise consolidations separated from the pleura and
328   larger airways. Numerous clinical conditions in which B-line artefacts can be present
329   additionally makes it difficult to recommend LUS as the sole diagnostic modality in cystic
330   fibrosis patients.

331

332 Authors have no conflict of interest

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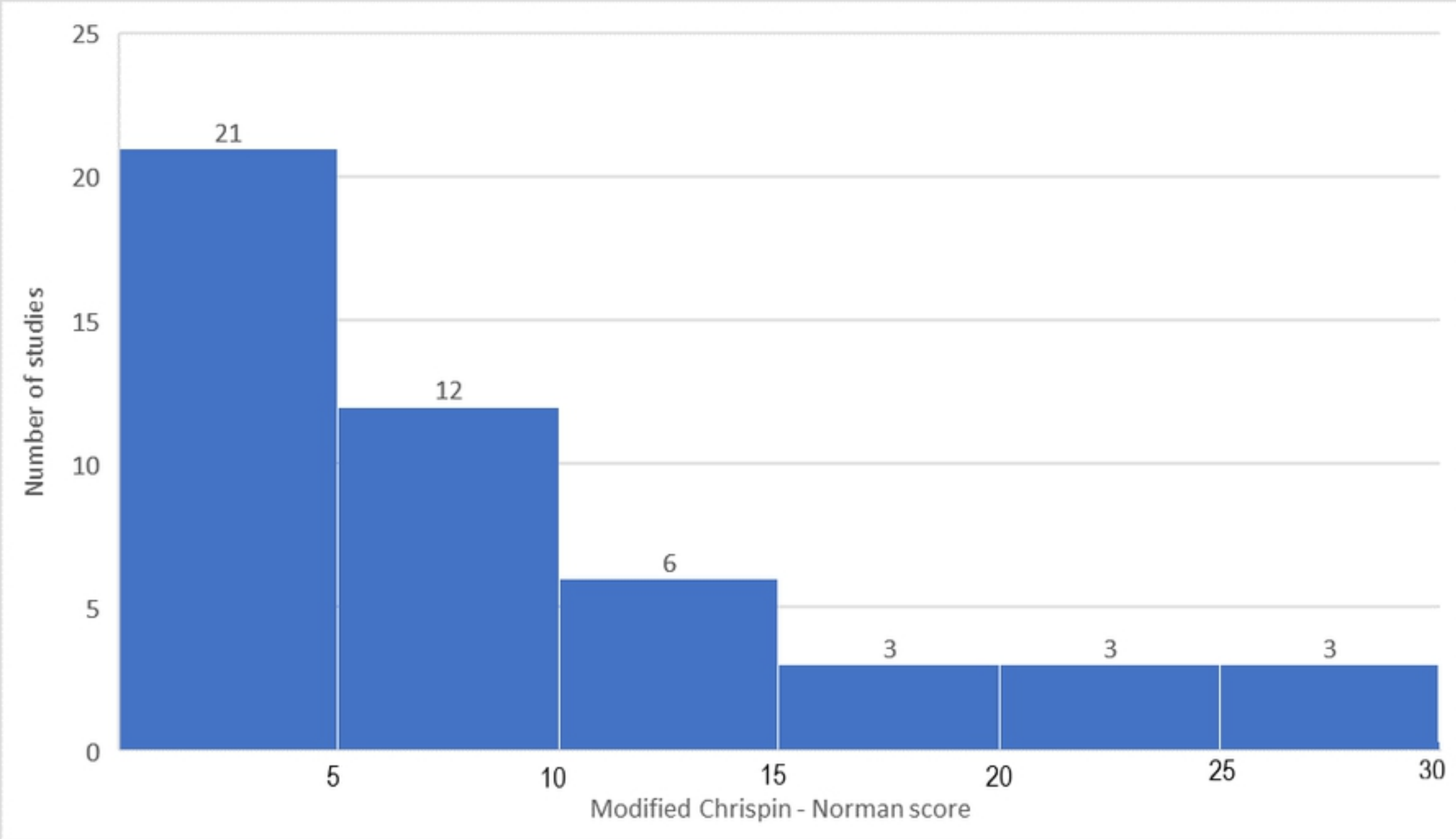
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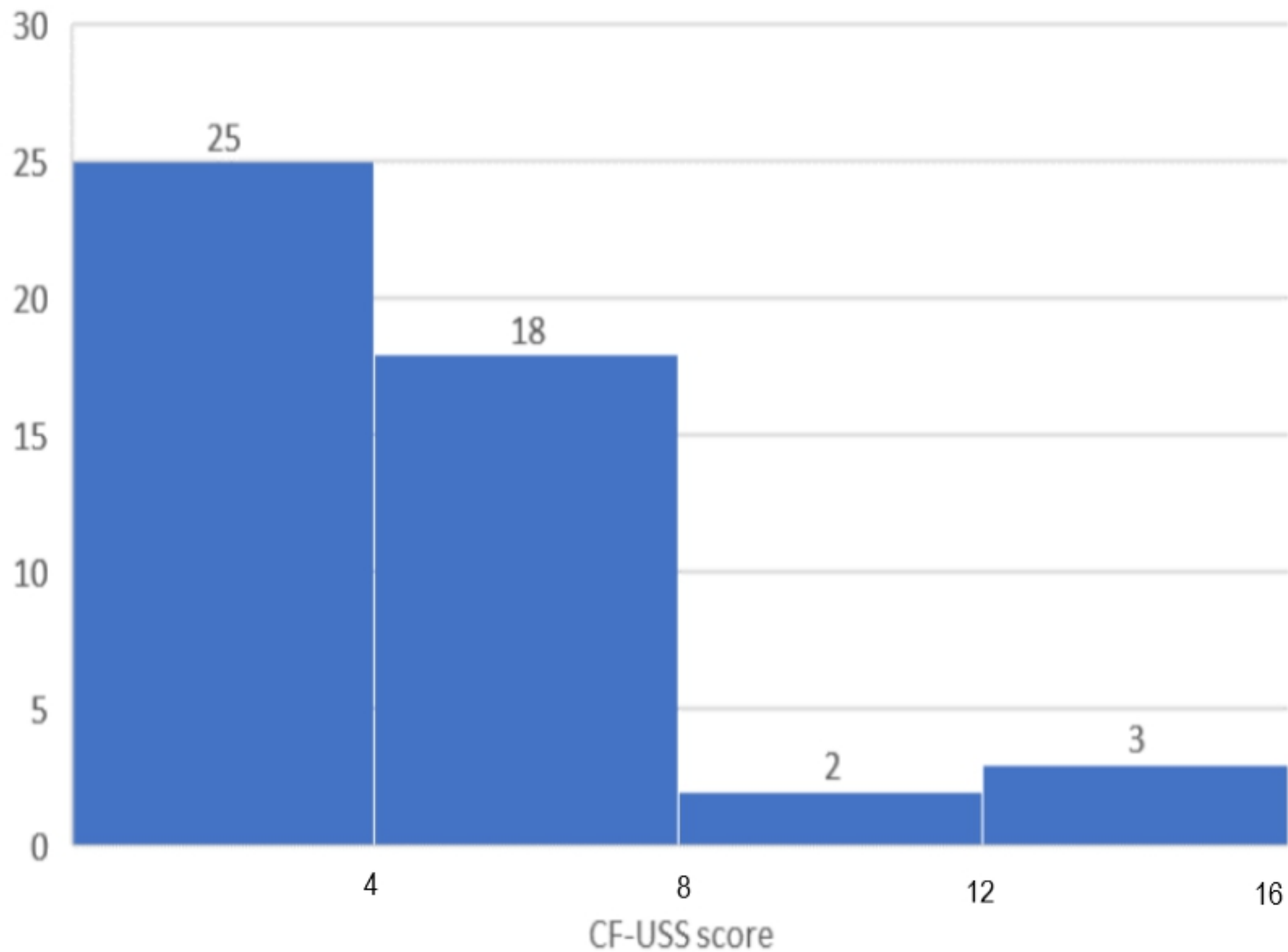
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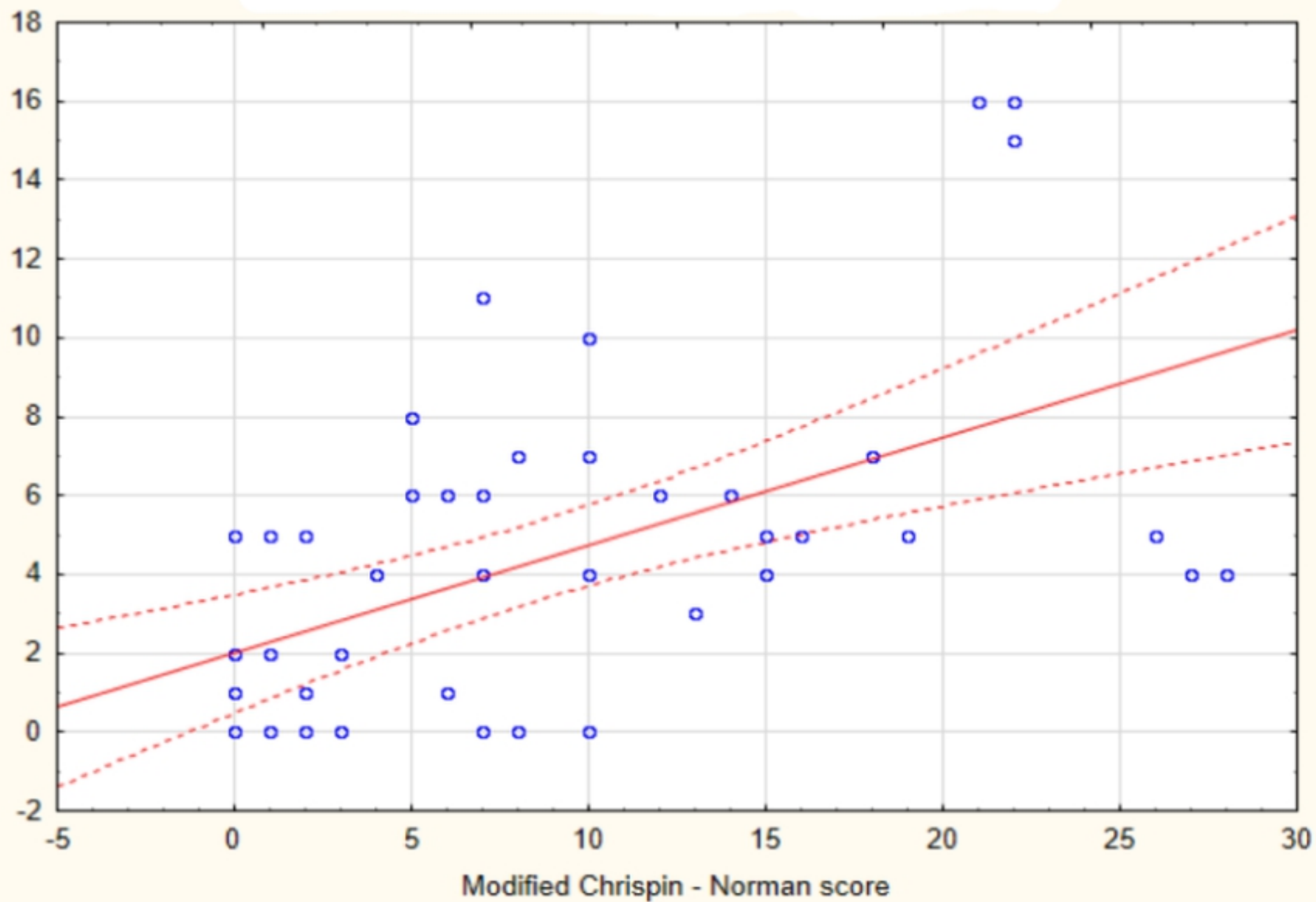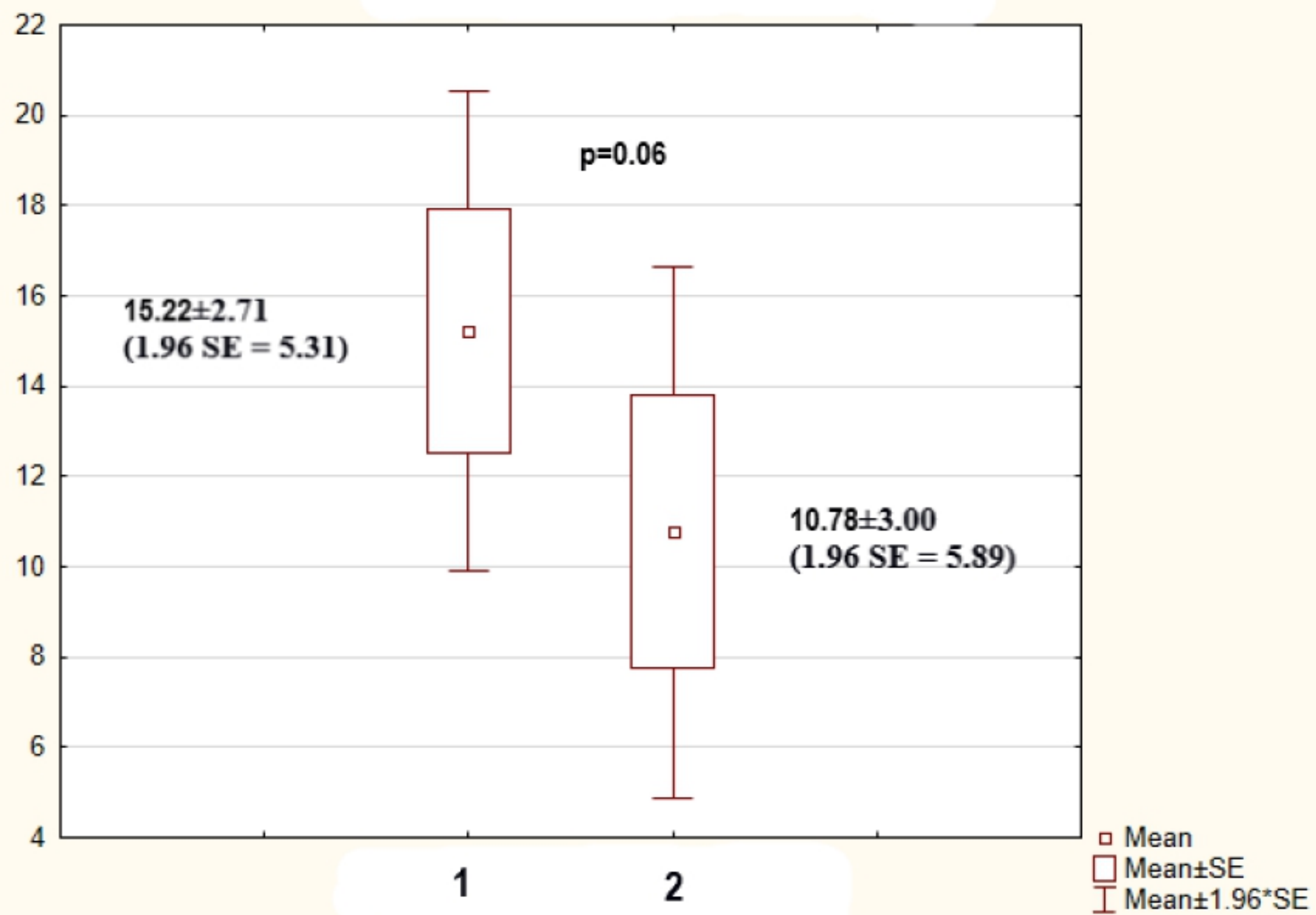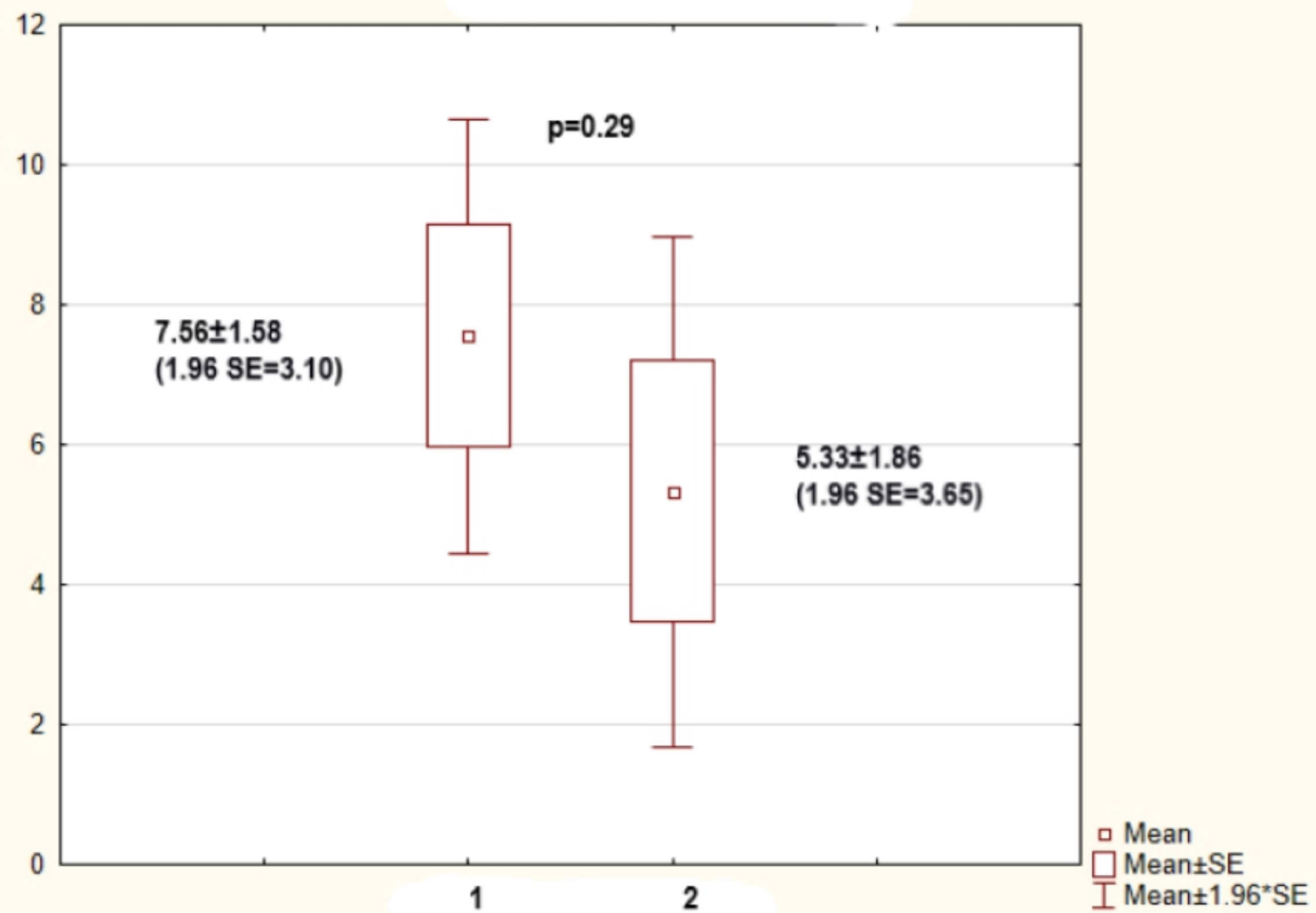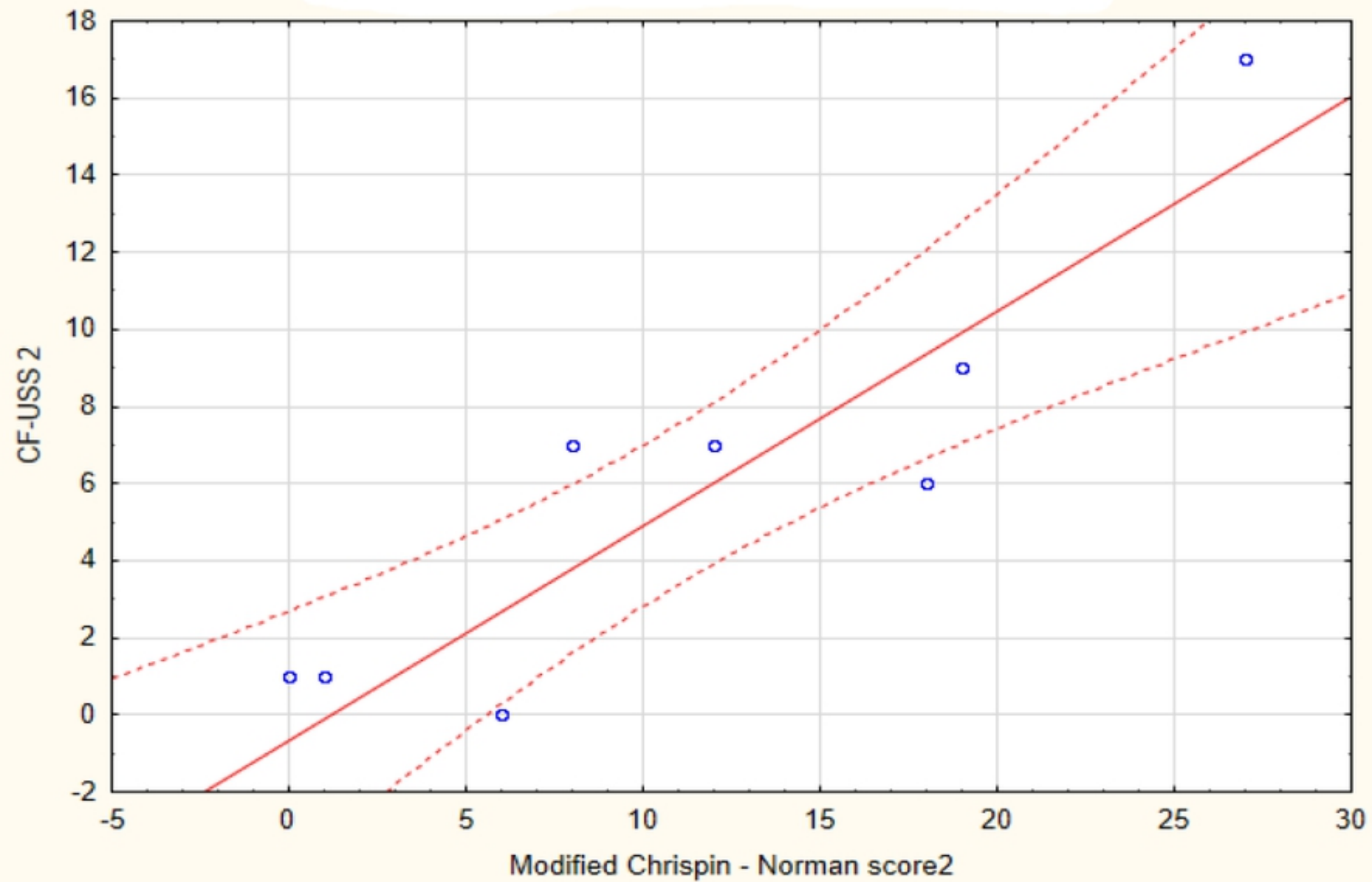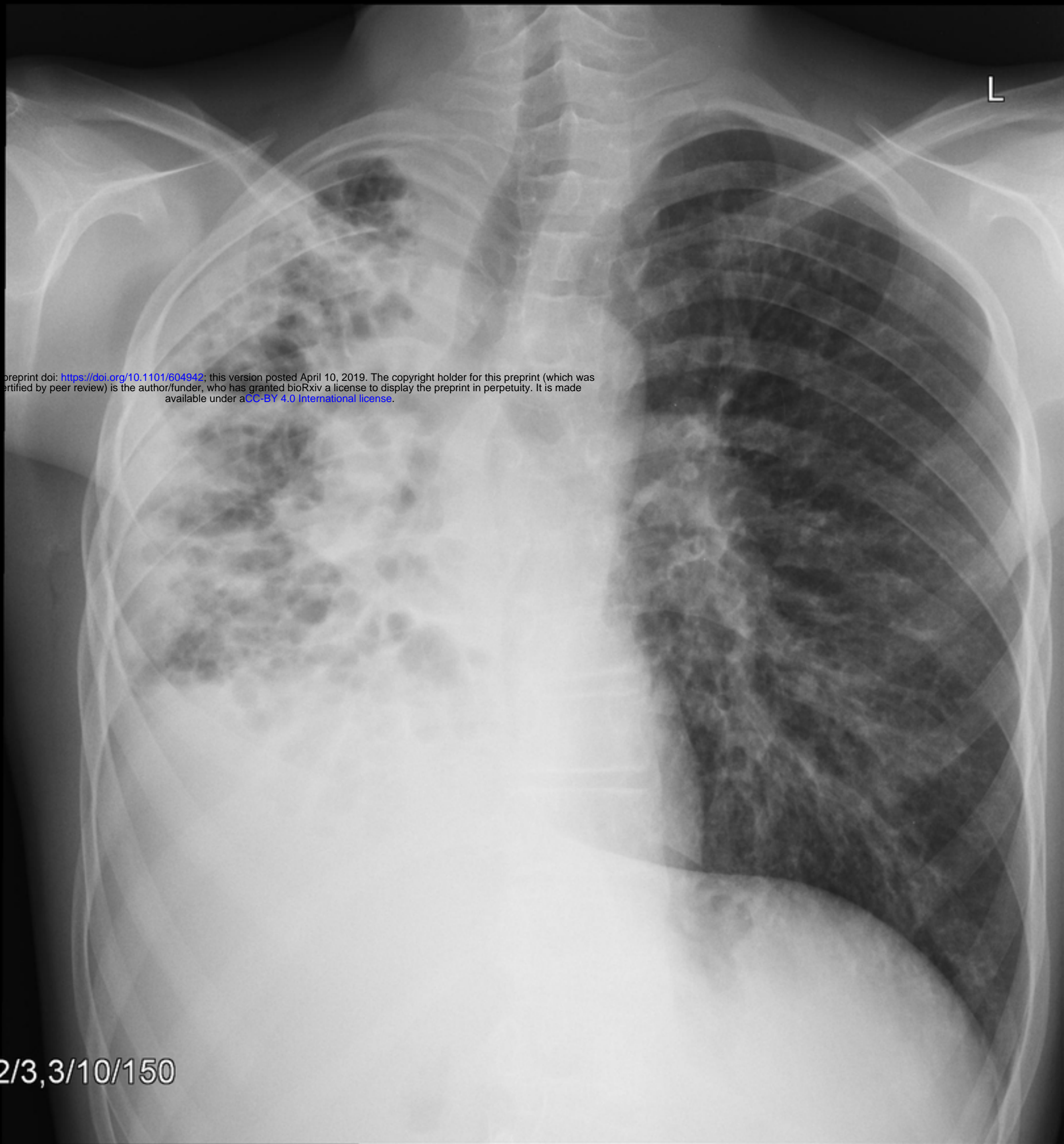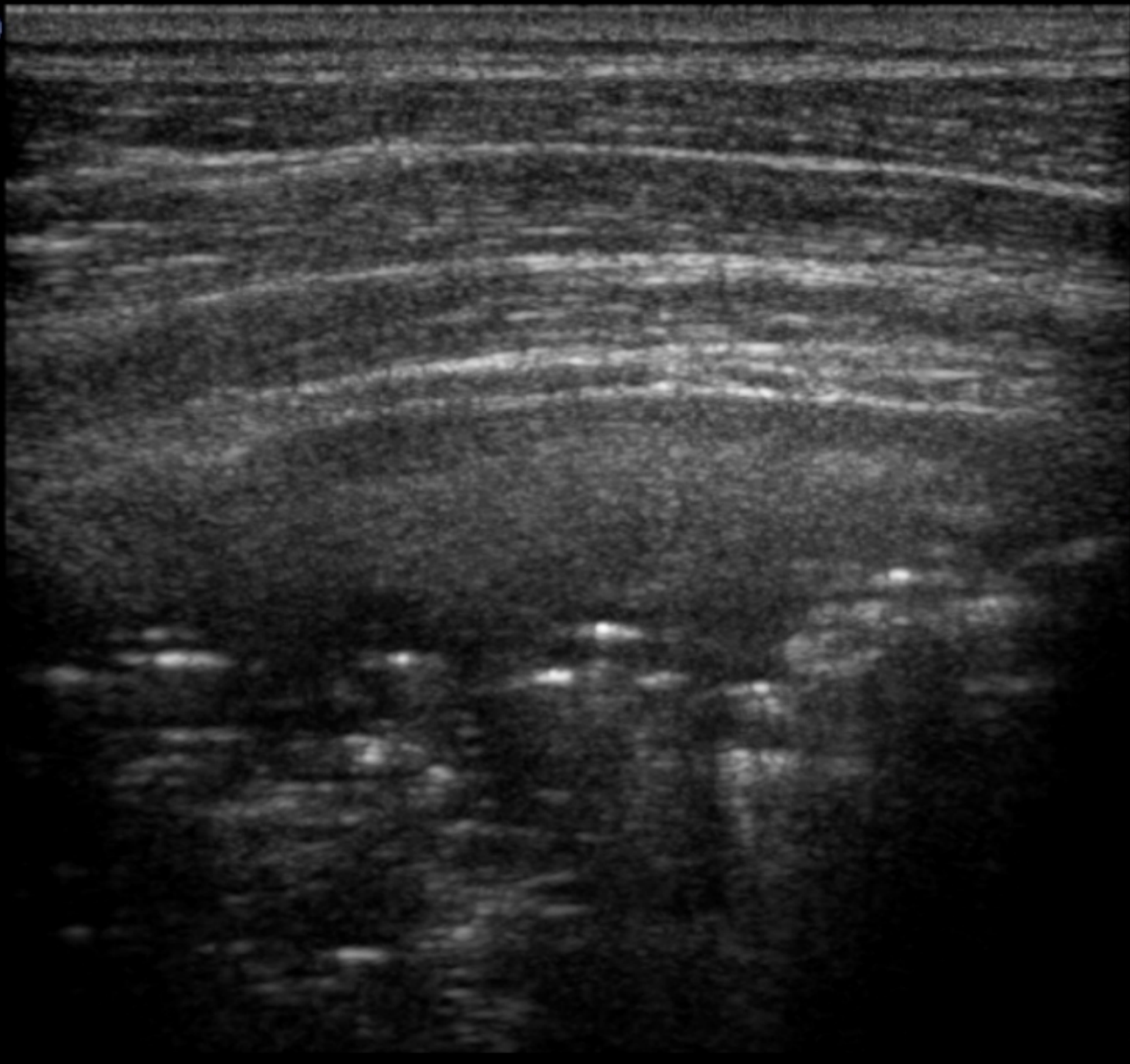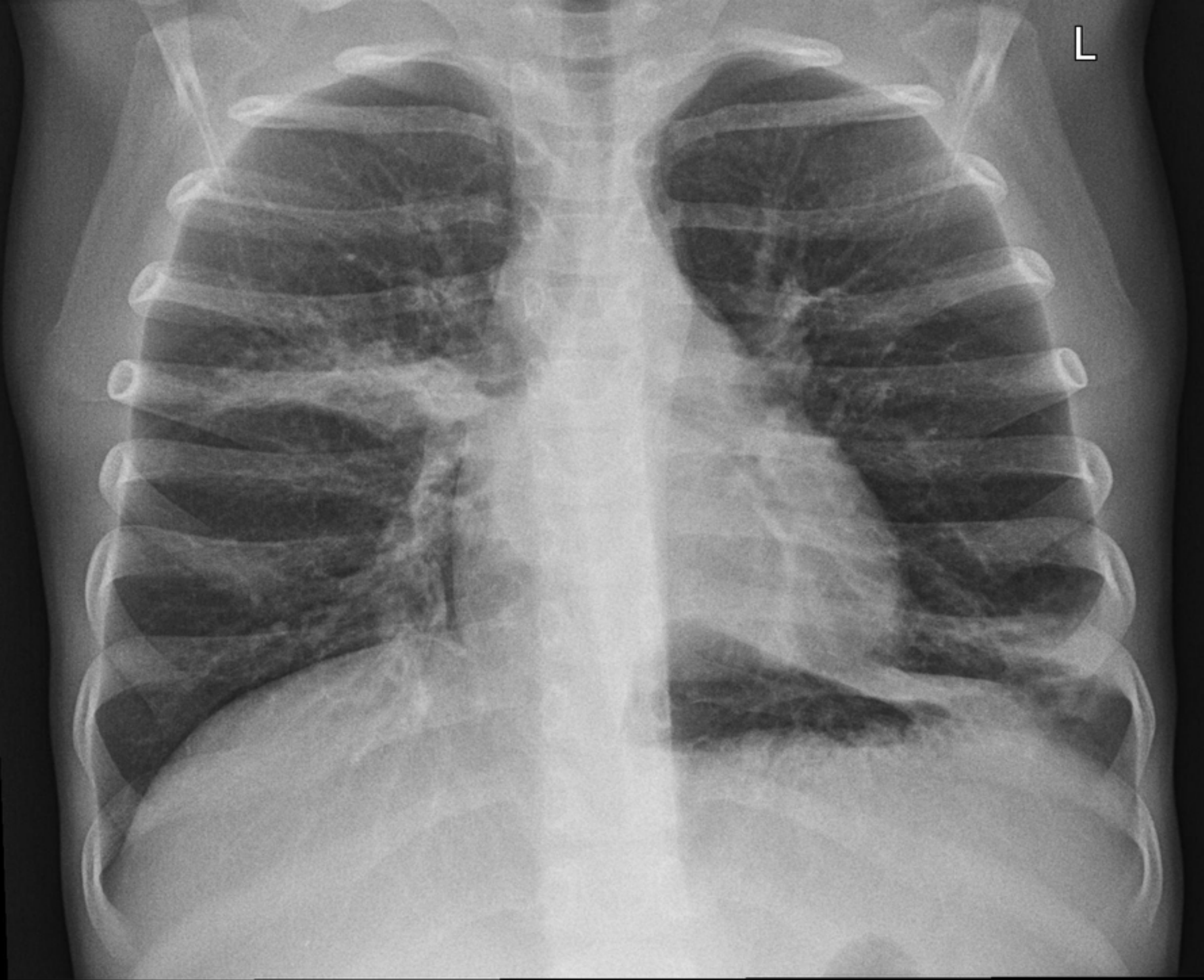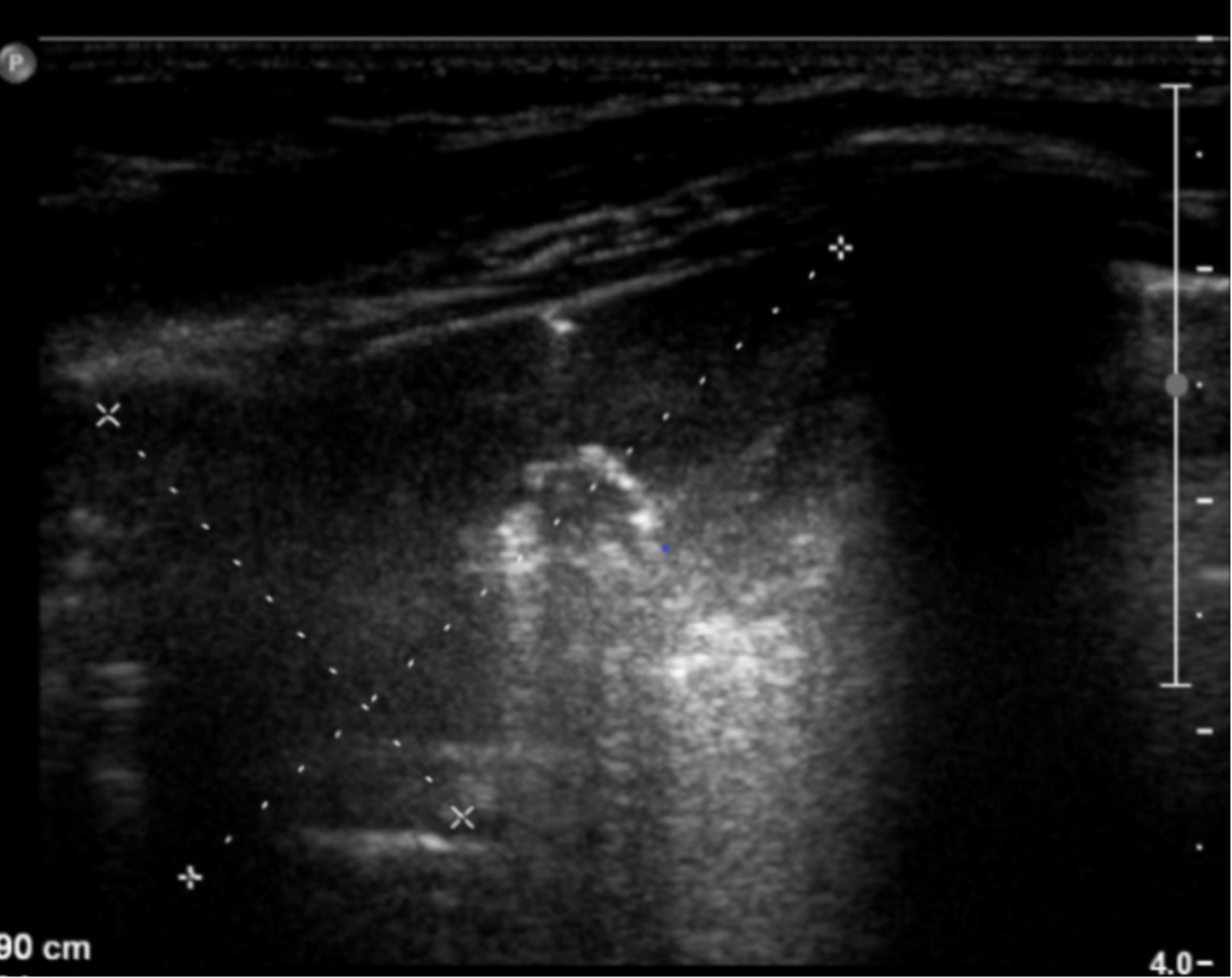
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