SYSTEMATIC REVIEW

Systematic Review of the Efficacy of Treatment for Median Arcuate Ligament Syndrome

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WHAT THIS PAPER ADDS

This systematic review suggests sustainable symptom relief above 70% after treatment for median arcuate ligament syndrome (MALS) in the majority of adult and paediatric studies, but the risk of bias is high and a formal meta-analysis could not be performed. This outcome supports guideline committees, acknowledging that MALS exists as a disease entity. However, studies of sufficient scientific quality are lacking to recommend specific treatments. To improve care for patients with MALS the next steps would be to deal with reporting standards, outcome definitions, and consensus descriptions of the intervention(s), after which an appropriate randomised controlled trial could be performed.

Objective: Since the first description of the median arcuate ligament syndrome (MALS), the existence for the syndrome and the efficacy of treatment for it have been questioned.

Methods: A systematic review conforming to the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) statement was conducted, with a broader view on treatment for MALS including any kind of coeliac artery release, coeliac plexus resection, and coeliac plexus blockage, irrespective of age. Online databases were used to identify papers published between 1963 and July 2021. The inclusion criteria were abdominal symptoms, proof of MALS on imaging, and articles reporting at least three patients. Primary outcomes were symptom relief and quality of life (QoL).

Results: Thirty-eight studies describing 880 adult patients and six studies describing 195 paediatric patients were included. The majority of the adult studies reported symptom relief of more than 70% from three to 228 months after treatment. Two adult studies showed an improved QoL after treatment. Half of the paediatric studies reported symptom relief of more than 70% from six to 62 months after laparoscopic coeliac artery release, and four studies reported an improved QoL. Thirty-five (92%) adult studies and five (83%) paediatric studies scored a high or unclear risk of bias for the majority of the Quality Assessment of Diagnostic Accuracy Studies 2 (QUADAS-2) items. The meaning of coeliac plexus resection or blockage could not be substantiated.

Conclusion: This systematic review suggests a sustainable symptom relief of more than 70% after treatment for MALS in the majority of adult and paediatric studies; however, owing to the heterogeneity of the inclusion criteria and outcome parameters, the risk of bias was high and a formal meta-analysis could not be performed. To improve care for patients with MALS the next steps would be to deal with reporting standards, outcome definitions, and consensus descriptions of the intervention(s), after which an appropriate randomised controlled trial should be performed.

Keywords: Coeliac Artery Compression Syndrome, Coeliac Artery Release, Dunbar Syndrome, Median Arcuate Ligament Syndrome, Plexus Blockage, Systematic Review

Article history: Received 27 February 2022, Accepted 28 August 2022, Available online XXX

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1078-5884/© 2022 Published by Elsevier B.V. on behalf of European Society for Vascular Surgery.
https://doi.org/10.1016/j.ejvs.2022.08.033

Please cite this article as: Metz FM et al., Systematic Review of the Efficacy of Treatment for Median Arcuate Ligament Syndrome, European Journal of Vascular and Endovascular Surgery, https://doi.org/10.1016/j.ejvs.2022.08.033

INTRODUCTION

The existence of median arcuate ligament syndrome (MALS) as a distinct entity caused by an increase of mucosal ischaemia,[1] and the efficacy of its treatment, has been questioned for a long time with respected physicians drawing different conclusions from the existing data.[2–4] The combination of a varying patient presentation, non-specific abdominal symptoms, and lack of validated, non-invasive mesenteric perfusion tests makes the diagnosis of MALS challenging to establish. Current international guidelines recommend that the diagnosis should be based on symptoms fitting chronic mesenteric ischaemia (CMI) and imaging studies showing compression of the coeliac artery (CA) by the median arcuate ligament (MAL) evaluated by an experienced multidisciplinary team, consisting of dedicated gastroenterologists, vascular surgeons, and radiologists. [2,3]

Ninety-six per cent of the experts on panel of the European CMI guidelines recommended that patients with MALS might be considered for surgical CA release, but consensus could not be reached on the first choice of treatment strategy and, consequently, clear recommendations are lacking.[3] Some publications have reported that local coeliac plexus blockage could be an alternative effective treatment for MALS,[5,6] but this has not been described as a treatment option in two comprehensive sets of guidelines.[2,3]

In 2012, Jimenez *et al.* published a systematic review of 20 retrospective studies reporting immediate symptom improvement in 85% of 400 patients with MALS after laparoscopic and open CA release, with a late recurrence in 19 patients in the open group (6.8%) and seven patients in the laparoscopic group (5.7%).[7] Limitations of the review by Jimenez *et al.* are that the evidence was based on mostly small individual series, the age of the patients was not reported, and the follow up period in the laparoscopic treatment group was short. After Jimenez *et al.*,[7] in 2020 Goodall *et al.* described more recent literature in a narrative review, but the review presented did not conform to the methodology of a systematic review. There has been no systematic review published on MALS treatment and outcomes in the past decade.[8] Furthermore, the most obvious outcome measure by today's standard, the impact of MALS on the quality of life (QoL) of patients before and after treatment, was not reported in the previous reviews.

The aim of the current systematic review was to summarise systematically the available literature on treatment, including open, laparoscopic, and robotic assisted CA release techniques, coeliac plexus blockage, and outcomes, including individual and societal gains in QoL, for adult and paediatric patients with MALS.

MATERIALS AND METHODS

Search strategy

The study protocol for this systematic review was registered with the international prospective register of systematic reviews (PROSPERO) and conformed to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement (PROSPERO CRD42021258592).[9] The literature search was performed according to the standards set out in the PRISMA statement.[9] Firstly, systematic searches in PubMed, the Cochrane Library, and Embase were performed to identify the relevant literature (details of the search strategy are shown in the Supplementary Literature Search). Secondly, all references cited in the existing reviews on MALS were hand searched for additional citations.

Study selection

The search focused on studies describing the outcomes MALS treatment published between its first description in 1963 up until 16 July 2021. Duplicates were removed. Patient cohorts could be included irrespective of age at presentation and treatment. The most important inclusion criteria were the presence of abdominal symptoms; proof of external compression of the CA on imaging; surgical treatment for MALS or plexus blockage; and at least three patients included in the study (Table 1). Grey literature was not included in this review.

Articles were independently selected by two authors (F.M.M. and J.T.M.B.), who were blinded to each other. The first selection was done by screening the titles and abstracts according to the pre-defined inclusion and exclusion criteria. Next, full text articles were read for inclusion in the final selection; consensus needed to be reached for an article to be included. In the case of disagreement, a third screener (R.H.G.) was involved to achieve consensus. Full texts were accessed via PubMed and through national and

Table 1. Inclusion and exclusion criteria of a systemic review of the treatment for median arcuate ligament syndrome (MALS)

Inclusion	Exclusion
Between 1963 up to 16 July 2021	Other than inclusion languages
English language	Comments, letter to editor, or other forms of own opinions without scientific substantiation
Randomised controlled trial, cohort, retrospective and prospective studies	Fewer than three patients included
External compression of the coeliac artery by the median arcuate ligament syndrome (MALS) on computed tomography angiography, magnetic resonance angiography, duplex ultrasound, or diagnostic angiography	No abstract or full text available
Abdominal symptoms	No treatment performed
Surgical treatment for MALS or plexus block	Surgery for MALS combined with other surgery
Outcomes of treatment reported	

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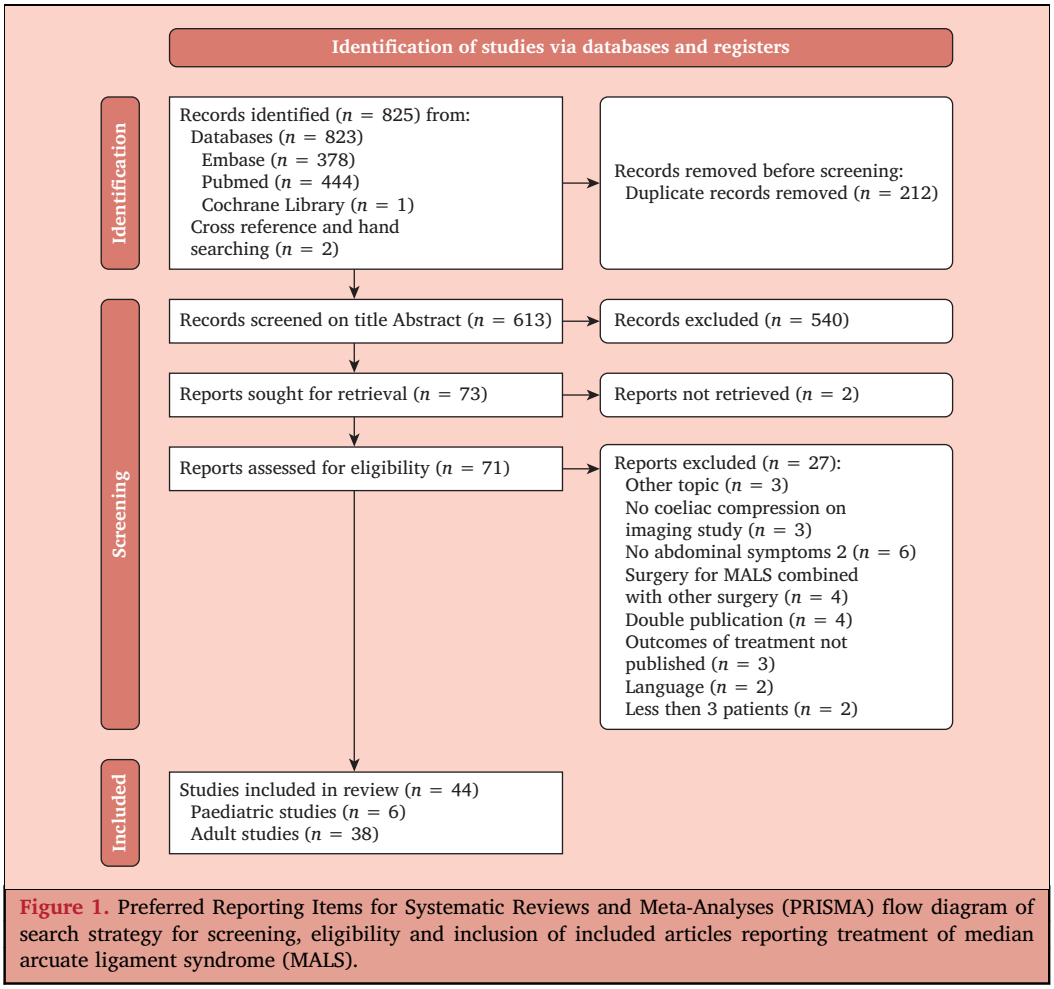

Figure 1. Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) flow diagram of search strategy for screening, eligibility and inclusion of included articles reporting treatment of median arcuate ligament syndrome (MALS).

international library requests. If full texts could not be retrieved, the article was excluded.

Outcome parameters

The following outcomes were described: study design and diagnostic criteria (imaging study, abdominal symptoms, and multidisciplinary diagnosis); Quality Assessment of Diagnostic Accuracy Studies 2 (QUADAS-2) appraisal of the included studies; and patient demographics (age, sex, duration of symptoms, body mass index, treatment, and additional procedures). The primary outcomes were symptom relief and QoL. The secondary outcomes were clinical outcomes (use of narcotics, death, complications, post-operative adjunctive procedures, psychiatric diagnosis, loss of productivity, and disability adjusted life years) and anatomical data (patency after treatment and duplex outcomes).

Continuous variables were displayed as means (with 95% confidence intervals [CIs]) or median (with interquartile range) for parametric and non-parametric data, respectively. Categorical variables were displayed as numbers (percentages). Patients aged less than 18 years and adults aged 18 years and older were analysed as separate cohorts (paediatric and adult, respectively).

All results were described in a narrative style. The primary and secondary outcomes described were divided according to the adult and paediatric literature. Data on symptom relief were presented as a forest plot to provide insight into the data at a study level.

Assessment of methodological quality

The risk of bias and applicability of the articles was evaluated by the QUADAS-2 tool.[10] The answers to the signalling questions and the applicability to the research were discussed after which the final appraisal was defined (F.M.M., J.T.M.B., and R.H.G.).

Data extraction

Data on study design, patient demographics, and outcome parameters were extracted from the included articles. Data on children (< 18 years old) were reported separately.

RESULTS

Search outcome and selection

A total of 611 papers were identified of which 58 including adults and 13 including paediatric patients (< 18 years old) were retrieved for full text review. Thirty-eight papers including adults and six including paediatric patients met the inclusion criteria and were ultimately selected for the final critical appraisal (Fig. 1).

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Table 2. Study design and diagnostic criteria for 880 adult patients as reported in 38 studies for the treatment of median arcuate ligament syndrome

Reference	Study design	Mean follow up − mo*	Lost to follow up − *n*	Patients − *n*	Imaging study	Abdominal symptoms >3 mo*	Multidisciplinary diagnosis
Baccari (2009)[42]	Retrospective	28		16	DUS and CTA or MRA	Yes	No
Barbon (2021)[5]	Prospective			22	CTA	Yes	No
Berard (2012)[33]	Retrospective	35		11	CTA	Yes, three mo unspecified	No
Berge (2020)[13]	Prospective	Median 18		12	CTA	Yes, three mo unspecified	Yes
Chaum (2021)[28]	Retrospective			4	DUS and CTA and MRA	Yes, three mo unspecified	No
Cienfuegos (2018)[12]	Retrospective	Median 117 (100−160)		11	CTA or MRA or DSA	Yes	Yes
Coelho (2020)[43]	Prospective	3		6	DUS or MRA	Yes	No
Columbo (2015)[23]	Retrospective	Median 7		21	DUS or CTA or DSA	Yes, three mo unspecified	No
De'Ath (2018)[31]	Prospective	Median 109 (78−114)		6	DUS and CTA or MRA or DSA	Yes, three mo unspecified	No
Do (2013)[24]	Retrospective			16	DUS or CTA or MRA or DSA	Yes, three mo unspecified	No
Dunbar (1965)[1]	Retrospective			13	DSA	Yes, three mo unspecified	No
Evans (1974)[22]	Retrospective			44	Unspecified	Yes, three mo unspecified	No
Fernstrum (2020)[34]	Retrospective	30	2	27	DUS and CTA	Unspecified	No
Geelkerken (2005)[17]	Retrospective	228	3	10	DSA	Yes	No
Grus (2018)[60]	Prospective	77		8	CTA	Yes	No
Ho (2017)[16]	Retrospective	Median 25 (6−72)	11	43	DUS or CTA or MRA or DSA	Yes	No
Kafadar (2021)[45]	Retrospective	6		10	CTA	Yes	No
Khrucharoen (2020)[32]	Retrospective	Median 16 (1−33)		41	DUS or CTA or MRA or DSA	Yes, three mo unspecified	No
Kohn (2011)[25]	Retrospective	49		6	Unspecified	Y, three mo unspecified	No
Marable (1968)[20]	Retrospective		2	19	DSA	Yes	No
Mihas (1977)[49]	Retrospective			4	DSA	Yes	No
Nguyen (2012)[61]	Retrospective	Median 15 (2−23)		5	DUS and CTA	Yes	No
Pather (2021)[41]	Retrospective	96 (48−144)	54	100	DUS or CTA or MRA	Yes	No
Reddy (2019)[50]	Retrospective	15		3	DUS	Yes	No
Reilly (1985)[26]	Retrospective	108	7	51	DSA	Yes, three mo unspecified	No
Rogers (1982)[19]	Retrospective	58	1	7	DSA	Yes, three mo unspecified	No
Roseborough (2009)[46]	Retrospective	Median 44		15	CTA or DSA	Yes	No
Sahm (2020)[47]	Prospective	5	9	18	DUS and CTA or MRA or DSA	Yes	No
Skelly (2018)[11]	Prospective	18	44	95	DUS and CTA or MRA or DSA	Yes, three mo unspecified	Yes
Sultan (2013)[27]	Retrospective	60		11	DUS and CTA	Yes, three mo unspecified	No
Takach (1996)[30]	Retrospective	44		7	DSA	Yes, three mo unspecified	No
Terpstra (1966)[18]	Retrospective	Median 12		5	DSA	Yes	No
Thoolen (2015)[14]	Retrospective	Median 6	1	9	DUS and CTA or MRA	Yes, three mo unspecified	Yes
Tulloch (2010)[48]	Retrospective	14		14	DUS or CTA or MRA or DSA	Yes	No

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Table 2-*continued*

Reference	Study design	Mean follow up − mo*	Lost to follow up − *n*	Patients − *n*	Imaging study	Abdominal symptoms >3 mo*	Multidisciplinary diagnosis
van Petersen (2017)[15]	Retrospective	6		129	DUS and DSA	Yes	Yes
Vaziri (2009)[29]	Retrospective	6		3	DUS and CTA and MRA	Yes, three mo unspecified	No
Watson (1977)[21]	Retrospective	Median 30 (14–33)	1	19	DSA	Yes	No
Weber (2016)[6]	Retrospective		14	39	DUS	Yes, three mo unspecified	Yes
Total	Retrospective: 31 Prospective: 7	Range 30–228	149	880			Yes: 6 No: 32

Data are presented as mean (95% confidence interval) or as median (interquartile range), unless stated otherwise. DUS = duplex ultrasound; CTA = computed tomography angiography; MRA = magnetic resonance angiography; DSA = digital subtraction angiography.
* Indicates whether the time scale of the symptoms has or has not been specified, and, if so, whether the patients experienced abdominal symptoms for more than three months.

Study design and diagnostic criteria

Adult cohort. The study design and diagnostic criteria of the 880 adult patients included are shown in Table 2.

The diagnosis of MALS was based on a consensus evaluation by a multidisciplinary team, as recommended in the international guidelines,[2,3] in only six of 38 adult studies.[6,11–15]

In nine studies, not all the reported adult patients (n = 280) met the inclusion criteria for the current review. As the data were reported for each individual patient, the results of the 223 individuals that did meet the criteria were included in this systematic review.[5,12,16–22]

In 18 adult studies, it was not clearly described whether patients had abdominal symptoms for at least three months. However, the fact that these patients received a comprehensive and time consuming evaluation and work up makes it likely that symptoms were present for more than three months.[1,6,11,13,14,19,22–33] In one study, symptom relief after treatment suggested that patients were symptomatic, although it was not clearly stated that patients did have abdominal symptoms at the time of inclusion.[34] Based on these considerations, these studies were included in the review.

In one adult study, one of 21 patients was a 16 year old,[23] and in another study two of 39 patients were 17 years of age. [6] The data of these young people could not be separated from those of the adults. Because of the low number (three of 60 patients, 5%) and the fact that they were adolescents, both studies are included in the text and tables covering adult patients.

Paediatric cohort. The study design and diagnostic criteria of the six included studies describing 195 patients aged < 18 years are provided in Table 3.[35–40]

In two of six studies, a diagnosis of MALS was based on a consensus evaluation by a multidisciplinary team;[38,40] one study did not describe the team members,[38] and in one study the team did not contain a radiologist,[40] as recommended in international guidelines.[2,3]

Quality Assessment of Diagnostic Accuracy Studies 2 (QUADAS-2)

Adult cohort. The QUADAS-2 appraisal of the studies that included adult patients is shown in Supplementary Table S1. Of note, 35 of 38 (92%) of the studies scored a high or unclear risk of bias for the majority of the items. No studies had a low risk of bias for all items. The study by Thoolen *et al.* scored a low risk of bias in all but one item (the index test), because they did not clearly define threshold values for the outcomes.[14] The study by van Petersen *et al.* scored high on the index test risk of bias and applicability, because they solely reported symptom relief and no asymptomatic patients.[15] The study by Weber *et al.* scored high on patient selection risk of bias because 14 of 39 patients were lost to follow up (LTFU).[6] They also scored high on the index test risk of bias and applicability, because they solely reported symptom relief and no asymptomatic patients, and they did not clearly define threshold values for the outcomes.

Paediatric cohort. In the paediatric cohort, five of the six studies (83%) scored a high or unclear risk of bias for the majority of the items of the QUADAS-2 appraisal (Supplementary Table S2). The sixth study scored high on patient selection risk of bias because 28 patients were LTFU in the QoL assessment. They scored high on index test risk of bias and applicability, because they solely reported symptom relief and no asymptomatic patients, and they did not clearly define threshold values for the outcomes.[38]

Patient demographics

Adult cohort. The demographics of the adult patients are reported in Table 4.

CA release was performed in 851 adult patients (97%) and the remaining 29 (3%) underwent a bypass, percutaneous CA stenting procedure, plexus blockage, or other operation on the CA.

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Table 3. Study design and diagnostic criteria for 195 paediatric patients as reported in six studies for the treatment of median arcuate ligament syndrome

Reference	Study design	Follow up − mo	Lost to follow up − *n*	Patients − *n*	Imaging study	Abdominal symptoms >3 mo*	Multidisciplinary diagnosis
Aschenbach (2011)[35]	Retrospective		0	22	MRA	Yes, duration unknown	No
Joyce (2014)[36]	Prospective	13 (2−24)		6	DUS and CTA or MRA	Yes	No†
Klimas (2015)[37]	Retrospective	Mean 62		58	DUS and CTA	Yes	No
Mak (2013)[38]	Prospective	9	28	46	DUS and CTA	Yes	Yes‡
Moak (2021)[39]	Prospective	22 (7−37)	1	31	DUS and CTA (and DSA if discrepancy)	Yes, duration unknown	No
Stiles-Shields (2018)[40]	Prospective	6		32	DUS and CTA or MRA or DSA	Unknown	Yes§
Total	Prospective: 4 Retrospective : 2	Range 6−62	29	195			Yes: 2 No: 4

Data are presented as mean (95% confidence interval), unless stated otherwise. MRA = magnetic resonance angiography; DUS = duplex ultrasound; CTA = computed tomography angiography; DSA = digital subtraction angiography.
* Indicates whether the time scale of the symptoms has or has not been specified, and, if so, if the patients experienced abdominal symptoms for more than three months.
† Consultation with vascular surgeon, paediatrician, and gastroenterologist but no team discussion
‡ No description of team members of the multidisciplinary team.
§ Multidisciplinary team without a radiologist.

Paediatric cohort. The demographics of the paediatric patients are reported in Table 6.

All patients in the paediatric cohort underwent laparoscopic MAL release.

Primary outcomes

Adult cohort. Symptom relief has been reported in Figure 2. Outcomes were not uniformly defined and reported. All studies reporting if patients were asymptomatic or experienced symptom relief after treatment were analysed. Some articles reported the number of patients who were asymptomatic after treatment, and some reported the number of patients who experienced a clear reduction of symptoms. These were reported separately (see Fig. 2). The forest plot was added to the table to provide a better insight into the data on a study level, without giving weight to one of the results. The error bars in the table represent the percentage of patients who experienced symptom relief, with the corresponding CIs. The percentages were calculated by dividing these by the number of patients in whom symptom relief was reported. Symptom relief was reported in 35 of the 38 adult studies describing 691 patients. In these 35 studies, 0% − 100% of the patients were free of symptoms and 75% − 100% reported a clear reduction of symptoms after surgical CA release. In 20 of 35 adult studies (57%), a symptom relief rate of > 70% was reported (range 71% − 100%; see Fig. 2).

Only one prospective adult study on plexus blockage met the inclusion criteria. This study showed symptom relief in 19 of 22 patients (86%).[5]

Five adult studies reported on QoL after treatment, as presented in Table 5. The prospective study by Skelly *et al.* was the only study to compare pre-operative with post-operative QoL, and revealed a statistically significant improvement in QoL from 68 to 80.3 ($p < .001$) on a visual analogue scale (VAS; 0 − 100) six months post-operatively.[11] The prospective study by Berge *et al.* reported on QoL 12 months post-operatively only in patients who experienced symptom relief.[13] The VAS scores of these nine patients improved from 44 pre-operatively to 62 post-operatively. In the EQ-5D-5L, four of the five dimensions improved. The retrospective study by Ho *et al.* showed a numerical difference in the 12 item Short Form Health Survey (SF-12) between surgically and conservatively treated patients with MALS, with a median follow up of 24 months.[16] In comparing the outcomes of the physical and mental domains of the SF-12, the surgical group did better (−5 [95% CI −17 − 10] *vs.* −9 [95% CI −22 − 4] and 1 [95% CI −6 − 8] *vs.* −9 [95% CI −19 − 2], respectively). The retrospective study by Pather *et al.* showed a significantly higher mean Gastrointestinal Quality of Life Index (GIQLI) of 80 (95% CI 3 − 97) in patients in whom the symptoms had disappeared compared with 53 (95% CI 38 − 68) in patients with persisting symptoms up to eight years post-operatively for MALS ($p < .001$).[41] The study by De'Ath *et al.* reported post-operative QoL scores only.[31]

Paediatric cohort. Symptom relief was reported in four of six studies ($n = 156$; Fig. 2); 77% − 98% of patients were free of symptoms and 63% − 67% reported a clear reduction in symptoms after laparoscopic CA release. The study by Aschenbach *et al.*[35] reported the number of patients who were free of symptoms and the number of patients who experienced symptom relief after treatment (both results presented in Fig. 2). Only the number of patients who

Table 4. Demographics of 880 adult patients as reported in 38 studies before treatment for median arcuate ligament syndrome

Reference	Female	Mean age − y	Mean duration symptoms − mo	Mean BMI − kg/m^2	Treatment	Additional peri-operative procedure − *n*
Baccari (2009)[42]	11 (69)	54	12	21	MALR	
Barbon (2021)[5]					Plexus block	
Berard (2012)[33]	9 (82)	Median 52	Median 41	20	MALR ($n = 10$), bypass ($n = 1$)	1
Berge (2020)[13]	8 (67)	46		21	MALR	
Chaum (2021)[28]	4 (100)	Median 30			MALR	2
Cienfuegos (2018)[12]	10 (91)	Median 34		21	MALR	
Coelho (2020)[43]	4 (67)	43			MALR	
Columbo (2015)[23]	16 (76)	Median 42		20	MALR	
De'Ath (2018)[31]	5 (83)	Median 30		Median 18	MALR	0
Do (2013)[24]	10 (63)		Median 16		MALR	
Dunbar (1965)[1]	12 (92)	34.5	Median 12		MALR	
Evans (1974)[22]					MALR	
Fernstrum (2020)[34]	18 (67)	49	57	27	MALR	
Geelkerken (2005)[17]		Median 47	24		MALR	
Grus (2018)[60]	5 (63)	61		21	MALR	8
Ho (2017)[16]	33 (77)	36			MALR	1
Kafadar (2021)[45]	6 (60)	42			MALR	
Khrucharoen (2020)[32]	37 (77)	Median 41		Median 21	MALR	
Kohn (2011)[25]	3 (50)	38			MALR	
Marable (1968)[20]	18 (95)	Median 37	Median 120		MALR	
Mihas (1977)[49]	4 (100)	48			Bypass ($n = 2$), other ($n = 2$)*	
Nguyen (2012)[61]		Median 29	26		MALR	
Pather (2021)[41]	75 (75)	38		23	MALR	25
Reddy (2019)[50]	2 (67)	39			MALR	
Reilly (1985)[26]	39 (76)	47	Median 12		MALR	18
Rogers (1982)[19]	6 (86)	44			MALR	
Roseborough (2009)[46]	13 (87)	Median 41			MALR	3
Sahm (2020)[47]	9 (50)	39	34	22	MALR ($n = 16$), bypass ($n = 3$)	
Skelly (2018)[11]	41 (80)	31	69		MALR	
Sultan (2013)[27]	10 (91)	50			MALR	3
Takach (1996)[30]	3 (43)	65			MALR	5
Terpstra (1966)[18]	4 (80)	Median 48			MALR	
Thoolen (2015)[14]	6 (67)	46	Median 12	Median 22	MALR	
Tulloch (2010)[48]	12 (86)	45			MALR	0
van Petersen (2017)[15]	106 (82)	35			MALR	
Vaziri (2009)[29]	3 (100)	44			MALR	
Watson (1977)[21]	11 (58)	Median 41			MALR	
Weber (2016)[6]	33 (85)	41		25	MALR	1
Total	568 (73)	Range 30−61	Range 12−120	Range 18−27	MALR ($n = 851$), bypass ($n = 3$), PTA ($n = 2$), plexus block ($n = 22$), other ($n = 2$)*	67

Data are presented as *n* (%) or mean (95% confidence interval), unless stated otherwise. BMI = body mass index; MALR = median arcuate ligament release; PTA = percutaneous transluminal angioplasty.
* Other operation on coeliac artery.

were free of symptoms (77%) was presented at the start of this section; presenting two sets of results for the same patients in this range could be misleading. In the studies in which patients experienced symptom relief (last sentence of this section), only the number of patients who experienced symptom relief (100%) is presented. In two of four paediatric studies (50%), symptom relief of > 70% was reported (range 98% − 100%; see Fig. 2).

Four paediatric studies reported an improvement in QoL after treatment (Table 6). The prospective study by Joyce *et al.* measured QoL via the Children Health Questionnaire − Parent Form 50 (CHQ-PF50), which reporting QoL

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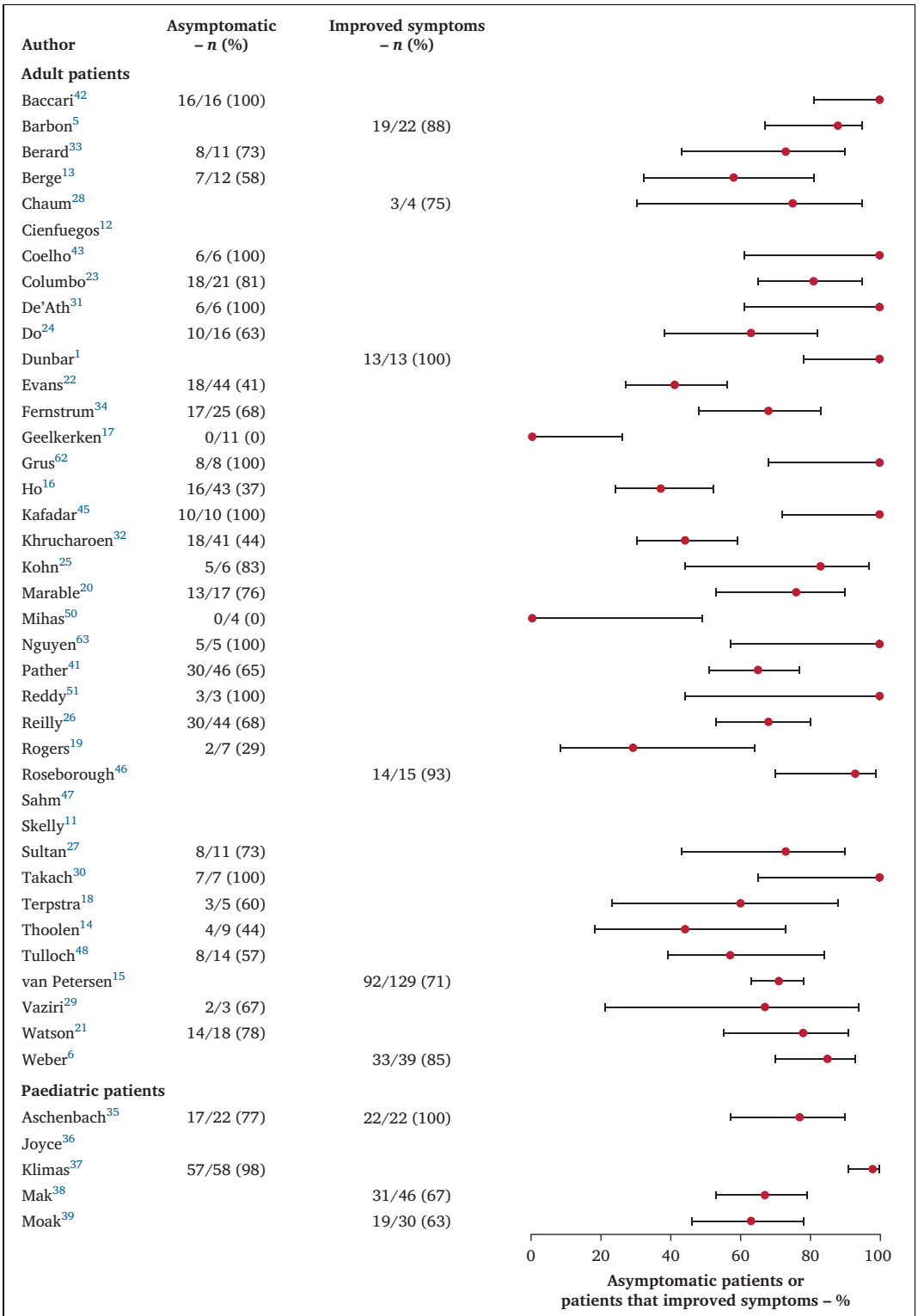

Figure 2. Symptom relief of patients among patients in whom the symptom relief was reported after treatment for median arcuate ligament syndrome as reported in a total of 43 studies. The error bars represent the percentage of patients who experienced symptomp relief with the corresponding confidence intervals.

according to seven domains reported by children and parents three months post-operatively.[36] The children reported a significantly improved score in "Physical Functioning", from 55 (20 — 90) to 96 (68 — 104) (P0.03); "Mental Health", from 42 (16 — 68) to 69 (48 — 89) (P0.03); and in "Self Esteem", from 47 (29 — 66) to 76 (60— 92) (P0.03). The prospective study by Mak *et al.* reported a significantly improved Pediatric Quality of Life Inventory (PedsQL) from 58 to 77 ($p < .001$) with a median follow up of 12 months.[38] The prospective study by Moak *et al.* reported

Table 5. Quality of life (QoL) of 880 adult patients after treatment for median arcuate ligament syndrome as reported in 38 studies

Reference	QoL pre-operative or conservative	QoL post-operative
Baccari (2009)[42]		
Barbon (2021)[5]		
Berard (2012)[33]		
Berge (2020)[13]	VAS 44	VAS 62
Chaum (2021)[28]		
Cienfuegos (2018)[12]		
Coelho (2020)[43]		
Columbo (2015)[23]		
De'Ath (2018)[31]		GLIQLI 129
Do (2013)[24]		
Dunbar (1965)[1]		
Evans (1974)[22]		
Fernstrum (2020)[34]		
Geelkerken (2005)[17]		
Grus (2018)[60]		
Ho (2017)[16]	Conservative SF-12: Physical −9 (−22–4); Mental −9 (−19–2)	SF-12: Physical −4.9 (−17–7); Mental 1 (−6–8)
Kafadar (2021)[45]		
Khrucharoen (2020)[32]		
Kohn (2011)[25]		
Marable (1968)[20]		
Mihas (1977)[49]		
Nguyen (2012)[61]		
Pather (2021)[41]		GLIQLI 71 (51–91)
Reddy (2019)[50]		
Reilly (1985)[26]		
Rogers (1982)[19]		
Roseborough (2009)[46]		
Sahm (2020)[47]		
Skelly (2018)[11]	68 (53–82)	80.3 (67–94)
Sultan (2013)[27]		
Takach (1996)[30]		
Terpstra (1966)[18]		
Thoolen (2015)[14]		
Tulloch (2010)[48]		
van Petersen (2017)[15]		
Vaziri (2009)[29]		
Watson (1977)[21]		
Weber (2016)[6]		

Data are presented as mean (95% confidence interval).
VAS = visual analogue scale; SF-12 = 12 item Short Form Health Survey; GLIQLI = Gastrointestinal Quality of Life Index.

self assessed QoL on a Likert scale from 1 to 10. QoL improved from 4.5 (2.4 — 6.6) to 5.3 (2.9 — 7.7; follow up QoL not described).[39] A prospective study by Stiles-Shields *et al.* reported a significantly improved PedsQL from 64 (47 — 80) to 74 (56 — 93; $p = .004$) six months post-operatively.[40]

Secondary outcomes

Adult cohort. A variety of secondary clinical outcome parameters have been reported in adult patients (Supplementary Table S3).

Twenty-one adult studies (including 512 patients) reported complications in 60 patients (12% of the patients); the most common was intra-operative bleeding in 24 patients (41% of the reported complications).[6,11,12,14,16,21,24,27,29–32,34,41–48] The "in hospital" and "30 days post-operative" mortality rate was zero, as reported in 21 studies including 368 patients.[5,6,18,19,21,23,24,27,29–33,41,43–46,48–50] In the study by Rogers *et al.*, one patient died two months after MAL release; post mortem examination failed to reveal the cause of death.

The anatomical outcomes of surgical CA release in studies including adult patients are provided in Supplementary Table 5. The adequacy of the CA release was determined with Doppler ultrasound in eight studies by reporting peak systolic velocity (PSV) values before and after surgery.[6,11,13,25,27,29,41,47] Two studies published pre- and post-operative inspiratory and expiratory PSV data;[41,47] one study only inspiratory values;[11] one study only expiratory values;[29] and in four studies it was not specified weither these were inspiratory or expiratory PSV data.[6,13,25,27] In nine studies including 274 patients, post-intervention CA patency was determined with duplex ultrasound, computed

Table 6. Demographics and quality of life (QoL) of 195 paediatric patients after laparoscopic median arcuate ligament release reported in six studies

Reference	Female*	Follow up − mo†	Age − y†	Pre-operative QoL†	Post-operative QoL†
Aschenbach (2011)[35]			15		
Joyce (2014)[36]	5 (83)	13 (2−24)	16 (14−17)	CHQ-PF-50 Physical Functioning 55 (20−90), Emotional 44 (6−83), Behavioural 59 (10 −108), Physical 26 (3−59), Bodily Pain 10 (−6−26), Mental Health 42 (16−68), Self-Esteem 47 (29−66), General Health Perceptions 17 (2−33)	CHQ-PF50 Physical Functioning 96 (68−104), Emotional 81 (49−113), Behavioural 83 (43−124), Physical 76 (37−115), Bodily Pain 57 (27−86), Mental Health 69 (48−89), Self-Esteem 76 (60−92), General Health Perceptions 48 (30−66)
Klimas (2015)[37]	47 (81)	Mean 62	17		
Mak (2013)[38]	42 (91)	9	16 (16−17)	PedsQL 58	PedsQL 77
Moak (2021)[39]	28 (90)	22 (7−37)	17 (15−19)	Likert 4.5 (2.4−6.6)	Likert 5.3 (2.9−7.7)
Stiles-Shields (2018)[40]	30 (94)	6	15 (14−17)	PedsQL 64 (47−80)	PedsQL 74 (56−93)
Total	152 (78)	Range 6−62	Range 15−17		

CHQ-PF50 = Child Health Questionnaire − Parent Form 50; PedsQL = Paediatric Quality of Life Inventory.
* *n* (%).
† Displayed as mean (95% confidence interval), unless stated otherwise.

tomography angiography, or magnetic resonance angiography.[6,15,26,28,33,42−45] A patent CA was established in 212 patients (77%).

Paediatric cohort. A variety of secondary clinical outcome parameters have been reported in paediatric patients (Supplementary Table S4). Because of the low patient numbers in which secondary outcomes have been reported, these will not be discussed further.

The anatomical outcomes of laparoscopic CA release in paediatric patients are shown in Supplementary Table S6). Doppler ultrasound was performed, and pre-operative and post-operative PSV values were reported in all six studies.

DISCUSSION

This systematic review, which included 38 studies describing the outcomes of 880 adult patients and six describing the outcomes of 195 paediatric patients, suggests a sustainable symptom relief above 70% in the majority of adults from three months up to 228 months after treatment for MALS and in half of the paediatric studies from six months up to 62 months after laparoscopic MAL release. Two adult studies compared QoL before and after surgical treatment for MALS and both showed an improved QoL after treatment. Four paediatric studies compared QoL before and after laparoscopic MAL release, and these also showed improved QoL after treatment. CA release was performed safely, with a very low complication rate and nearly zero probability of death.

Only one study, including 22 adult patients, reported symptom relief of 88% after coeliac plexus blockage.[5]

None of the articles included in the present review was of sufficient quality to meet the criteria for a "low risk" score, according to the QUADAS-2 tool. Most importantly, in the majority of studies, outcome parameters were ill defined and not uniformly defined and or presented, with a risk of confounding and selection bias. The consequence is that the results must be interpreted with caution.

When starting this review, the idea was to perform a systematic review with comparative meta-analysis of the data. The outcome of the previous reviews allowed for both a systematic and a scoping review approach. A systematic review design was chosen in line with the recommendation in the most recent guideline describing diagnostic criteria.[3] Based on the design of the included articles (solely observational studies and case series), the "quality of evidence" of all studies would be "low" or "very low".[51] Owing to the heterogeneity of the inclusion and outcome criteria, it was decided to perform solely a narrative description of the results without a formal meta-analysis, which would provide the readers with misleading data that should not be (mis)used in future reports. The justified criticism of performing a systematic review in a rare disease is the risk of selection and publication bias. After exploring the data, the actual quality of evidence appeared to be very low due to the observational study designs, inherent patient selection, and variable outcome parameters. Without data synthesis, a GRADE (Grading of Recommendations, Assessment, Development and Evaluations) evaluation could not be completed. To answer the research question, both guidelines committees recommended to perform a randomised controlled trial.[2,3]

The reported symptom relief above 70%, as presented in the majority of the studies, is substantially higher than the placebo response of 18% − 57% described in five sham operation studies.[52−56] Jimenez *et al.* presented an 85% immediate symptom improvement of 400 patients with MALS after laparoscopic and open CA release with a late recurrence

Please cite this article as: Metz FM et al., Systematic Review of the Efficacy of Treatment for Median Arcuate Ligament Syndrome, European Journal of Vascular and Endovascular Surgery, https://doi.org/10.1016/j.ejvs.2022.08.033

in 19 patients in the open group (6.8%) and seven patients in the laparoscopic group (5.7%).[7] Owing to the above arguments, it is inappropriate to present aggregated data on these studies. After Jimenez *et al.*, Goodall *et al.* carried out a literature overview in 2020, which was presented as a description of some of the literature on MALS without a methodology or the design of a systematic review.[8] The current review included 44 studies of adult and paediatric patients (1 096 patients in total), 17 of which (266 patients) were also included in the study by Jimenez together with 27 new studies (830 patients). The second difference between the present work and the previous studies is that the articles included in the current research had a longer follow up period of three months up to 228 months, compared with the articles included by Jimenez *et al.* A third difference is that the current review applied more stringent selection criteria. This systematic review only included studies if patients had external compression of the CA by the MAL on imaging studies plus abdominal symptoms (for more than three months), whereas Jimenez *et al.* included all studies that presented outcomes after surgical treatment for MALS.[7]

A limitation of this review is that the aim was to only include patients with abdominal symptoms, but some articles were included based on the fact that it was very likely that they experienced abdominal symptoms for at least three months.[1,6,11,13,14,19,22–34]

This systematic review included a separate description of paediatric studies showing sustainable symptom relief rate of 63% – 98% after laparoscopic MAL release. The present review underlined that MALS in patients younger than 12 years of age is rare, supporting the hypothesis that compression of the CA arises during puberty as the thorax/abdominal ratio increases.

Two prospective cohort studies provided evidence that patients with MALS have ischaemia of the gastric mucosa and showed improvement of validated mesenteric mucosal perfusion tests after successful CA release.[13,57] Moreover, in a study of 129 patients, 91 (71%) experienced relief of symptoms, irrespective of the fact that the coeliac plexus had been left untouched during surgery.[15] This does not necessarily mean that the ischaemia is the cause of the pain, nor that neurogenic mechanisms are also involved. The present systematic review undermines the half century old statement of Szilagyi *et al.*[4] that "no patient had ever been proven, on scientific grounds, to have an abnormality of intestinal structure or function which was caused by extraluminal compression of the coeliac artery, or supposed relief from the operation could be anything other than a placebo effect". The present review supports both guideline committees, acknowledging not only that MALS exists as disease entity, but also that studies of sufficient scientific quality are lacking to recommend specific treatments.[2,3] To facilitate the development of evidence based guidelines for the management of MALS, both guideline committees recommend a blinded RCT comparing CA release with a sham operation. A systemic review reconsidering the ethics of sham interventions concluded that sham interventions are acceptable, provided the pre-conditions of scientific necessity, reasonable risks, and valid informed consent are fulfilled.[58] The suggestion of a randomised placebo controlled patient and observer blinded clinical superiority trial in patients suspected of MALS will give the irrefutable answer needed by patients and clinicians. Included patients should have a multidisciplinary consensus diagnosis based on symptoms fitting CMI, combined with appropriate imaging studies,[59] and should be treated by either surgical CA release or a sham operation. Besides analysing anatomical and clinical success, the outcome parameters should represent QoL, psychiatric disorders, and the societal burden of disease. If these criteria are met, the Szilagyi debate may finally be settled, and it will either underline the usefulness of surgical CA release as a (cost) effective treatment for MALS or it will prevent patients with disabling abdominal complaints from undergoing an ineffective intervention.

In conclusion, this systematic review suggests a sustainable symptom relief above 70% after treatment for MALS in the majority of the adult and paediatric studies, but, owing to the heterogeneity of the inclusion criteria and outcome parameters, the risk of bias is high and a formal meta-analysis could not be performed.

CONFLICT OF INTEREST STATEMENT AND FUNDING

None.

APPENDIX A. SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ejvs.2022.08.033.

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Please cite this article as: Metz FM et al., Systematic Review of the Efficacy of Treatment for Median Arcuate Ligament Syndrome, European Journal of Vascular and Endovascular Surgery, https://doi.org/10.1016/j.ejvs.2022.08.033

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Please cite this article as: Metz FM et al., Systematic Review of the Efficacy of Treatment for Median Arcuate Ligament Syndrome, European Journal of Vascular and Endovascular Surgery, https://doi.org/10.1016/j.ejvs.2022.08.033