# A Systematic Review of Primary Rhinoplasty in Patients With Bilateral Cleft Lip

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**Background:** Primary rhinoplasty (PR)at the time of cleft lip repair is controversial. We previously performed a systematic review that supported PR during unilateral cleft lip repair. We now aim to determine whether the same idea translates to care of patients with bilateral cleft lip.

**Methods:** A systematic review was conducted adhering to Preferred Reporting Items for Systematic Reviews and Meta-analysis statement guidelines. PubMed and Embase databases were searched for studies that met our inclusion criteria: (1) English language, (2) human subjects, (3) rhinoplasty at the time of bilateral cleft lip repair, and (4) evaluation of nasal outcome. Studies were excluded in case of: (1) inclusion of a large proportion of syndromic patients, (2) case reports, (3) editorials, (4) letters, (5) reviews, and (6) exclusive to unilateral clefts. Out of 281 studies that showed up on initial search, 12 were included in our review. Research quality and level of evidence rating were determined for each study.

**Results:** Of the 12 included studies, 9 supported PR at the time of bilateral cleft lip repair; 8 studies evaluated nasal growth and found no restriction over time; 4 studies followed 158 patients to an average of 15 years and showed 77% did not need secondary rhinoplasty.

**Conclusion:** Although the available literature supports PR in patients with bilateral cleft lip with respect to subjective and objective outcomes, nasal growth, and reducing the need for secondary/revision rhinoplasty, there are significant limitations, necessitating large volume studies.

**Key Words:** bilateral cleft; cleft lip; cleft nasal deformity; primary cleft lip nose repair;, primary rhinoplasty

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The worldwide reported incidence of cleft lip varies from 0.2 to 2.3 per 1000 births, of which, 10% to 25% have a bilateral cleft.[1] Patients with bilateral cleft lip have a complex nasal deformity that tends to be exacerbated with the growth of the child if not addressed surgically.[2] The appropriate approach to correct this deformity has long been debated with respect to timing, technique, and extent of intervention.[3]

In 1967, Millard pioneered the idea of simultaneously repairing the bilateral cleft lip with the nasal deformity in a single operation.[4] This technique, however, was met with a great controversy over the potential impairment of nasal growth and has since remained a topic of debate. In 1985, McComb published a 10-year longitudinal study that demonstrated promising results with primary correction of the nasal deformity in patients with unilateral cleft lip.[5] This study transformed cleft care as it proposed that primary rhinoplasty (PR) during cleft lip repair did not disrupt nasal growth. These encouraging reports, among others[6–8] led to the resurgence of PR within cleft lip repair, which has since become the standard of care. The debate, however, continues on whether and to what extent PR should be performed, its effect on nasal growth, and whether it eliminates the need for further rhinoplasty at skeletal maturity in patients with bilateral cleft lip. To address this, we designed this study to collect, organize, and interpret the available data in the literature. We have previously published a systematic review on the topic of PR in unilateral cleft lip nasal deformity.[9]

The aim of our study is to determine the amount and quality of evidence for and against primary bilateral cleft rhinoplasty, how it affects nasal growth, and its effect on reducing the proportion of patients needing subsequent revision rhinoplasty. We will also compare the surgical techniques for rhinoplasty described in the literature.

## METHODS

We designed a systematic review after the guidelines established by the Preferred Reporting Items for Systematic Reviews and Meta-analysis statement. PubMed was utilized to search the US National Library of Medicine (NLM) database in January 2020 using the terms: "Bilateral Cleft Rhinoplasty" and "Primary Cleft Lip Nose Repair". A similar search was done utilizing Embase in October 2021 using the term: "Bilateral Cleft Rhinoplasty; Nasal". No constraints were placed on the study setting, surgical approach, evaluation of outcomes, or year of publication. To be included, the studies needed to have human subjects undergoing rhinoplasty at the time of bilateral cleft lip repair, and some evaluation of the nasal outcome. Exclusion criteria included: (1) inclusion of a large proportion of syndromic patients, (2) case reports, (3) editorials, (4) letters, (5) reviews, and (6) studies exclusive to unilateral clefts. The full text of the included studies were screened by 2 independent

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investigators. The input of the principal investigator was utilized when there was a dissonance of opinions.

The data extracted from the articles were charted in a Microsoft Excel (Version 16.58) spreadsheet. It considered study design, number of patients included, technique of PR, outcome assessment type (subjective/ objective), average follow-up time, presence of control group, outcome measures used, percentage of patients not needing secondary rhinoplasty, and effect on nasal development over time. Statistical analyses were conducted on Microsoft Excel. Level of evidence was determined using American Society of Plastic Surgeons (ASPS) Evidence Rating scale and research quality was assessed using Cochrane's Risk Of Bias In Non-Randomized Studies- of Intervention (ROBINS-I) tool for cohort studies. All Preferred Reporting Items for Systematic Reviews and Meta-analysis systematic review guidelines were followed including checklist, flowchart, and article tables.

## RESULTS

A total of 165 and 116 English language articles were identified from the initial search queries on PubMed and Embase, respectively. We screened 281 titles and abstracts to see whether they met our inclusion criteria. Sixty-four abstracts met the criteria of which 52 full-text articles were excluded. In total, 12 studies (9 from PubMed and 3 from EMBASE) were included in our systematic review. Full breakdown of studies included and excluded can be seen in Figure 1.

Of the 12 articles reviewed, 5 were retrospective cohort studies, 5 were case series, and 2 were single-surgeon case series. In total there were 462 bilateral cleft lip patients, with the smallest study consisting of 7 patients[10] and the largest 109 patients.[11] Studies were published between 1986 and 2021. There was significant variation in the rhinoplasty techniques, none of which included a septoplasty.

The study design, number of patients and rhinoplasty technique used for each of the 12 studies is outlined in Supplemental Table 1, Supplemental Digital Content 1, http://links.lww.com/SCS/E256, and the outcome assessment is presented in Supplemental Table 2, Supplemental Digital Content 2, http://links.lww.com/SCS/E257.

### Article Level of Evidence and Risk of Bias

The highest ASPS level of evidence rating for any included studies was Therapeutic III (T III). Five studies had level of evidence rating T III whereas 4 were rated T IV level of evidence. Cochrane's Risk of Bias in Non-Randomized Studies of Interventions (ROBINS-I) tool was used to assess the research quality of the 5 retrospective cohort studies in our systematic review.[12] We determined the risk of bias as critical for 2, serious for 2, and moderate for 1 of these studies. Figure 2 illustrates the risk of bias assessment using a traffic light plot (created using Cochrane's robvis tool).

### Subjective Outcomes

The results of rhinoplasty were assessed subjectively in 5 papers[7,11,13–15] in multiple ways, including 5-point Visual Analog Scale (VAS),[14] subjectively by the author[7,11,13–15] and the surgical team along with laypersons.[14] Three studies[7,14,15] used both subjective and objective ways of assessing outcome.

Three of the studies did not include a comparison group[7,13,15]; however, the other 2 did. One study compared results with patients who underwent unilateral cleft lip repair[11]; whereas the other compared different surgical techniques—PR alone, naso-alveolar molding (NAM) alone, NAM plus PR, NAM plus PR with overcorrection- with a control group of cleft palate patients that underwent palatoplasty.[14]

Of the 3 studies that did not utilize a comparison group, 2 presented positive results in their subjective assessments during follow-up. One found that 75% of patients had normal nostril shape and symmetry[15] whereas the other found effective and stable repair with no growth interference or noticeable external nasal scars.[13] The single study that showed unsatisfactory subjective results found an increase in columella length after 15 years leading to larger nostrils, broadening of the nasal tip, and downward drift of the columellar base.[7]

The 2 studies that used comparison groups in determining subjective outcomes gave mixed results. One found positive long-term outcomes with respect to speech, occlusion, and facial balance; however, when compared with results of unilateral cleft patients, the long-term outcome of lip and nose were worse.[11] This was determined in the senior author's opinion but not necessarily rigorously determined. The other study found that patients who underwent NAM plus PR with overcorrection had a nasal appearance most similar to patients with cleft palate who underwent palatoplasty alone. These patients also had the highest 5 point Visual Analog Scale (VAS) score of 4.59/5.[14]

### Author Satisfaction Outcomes

Of the 2 studies that discussed outcomes with regards to author satisfaction,[6,10] both supported PR with bilateral cleft repair. One such study found that only 1 of the 33 patients included in the case series required additional surgery at 10 years after the procedure.[6]

### Objective Outcomes

Eight publications measured results objectively.[7,14–20] Assessment included 2D indirect anthropometric measurements,[14,15,17] 3D indirect anthropometric measurements,[16,20] direct anthropometric measurements,[19] serial casting for measuring anthropometric distances,[18,19] and comparison of columellar length.[7,18,19]

All but one of these studies used age-matched norms as comparison groups.[7,15–20] One of the studies compared different PR techniques in bilateral cleft lip repair with cleft palate patients who underwent palatoplasty.[14] Another study compared PR with a 2-stage lip and nasal repair.[19] Another study compared 2 different techniques of PR (Trott and Mulliken types) with age-matched normal controls.[17]

Seven (87.5%) of the publications with objective evaluation supported PR during cleft repair. Of the 3 articles that used 2D indirect anthropometric measurements, 1 identified a concern with PR.[17] Of the 2 that supported PR during cleft repair, 1 found no significant difference in columella length between cleft patients and age-matched controls[15] and the other found that NAM plus PR with overcorrection resulted in a nasal appearance most similar to their control group of cleft patients who had undergone palatoplasty only.[14] The article that identified a concern noted that many nasal parameters were similar to age-matched controls, but both techniques resulted in increased nasal width, nasolabial angle, and nasal angle; the columellar length and nasal tip projection was shorter in the Trott group compared to Mulliken.[17]

3D indirect anthropometric measurements using age-matched controls found no differences in linear, angular, and proportional dimensions of the nose,[16,20] although 77% of patients underwent an intermediate procedure before age 6 to elongate the columella, reposition the lower lateral cartilages, and improve tip projection.[16] Direct anthropometric measurements revealed that the measurements of nasolabial anatomy

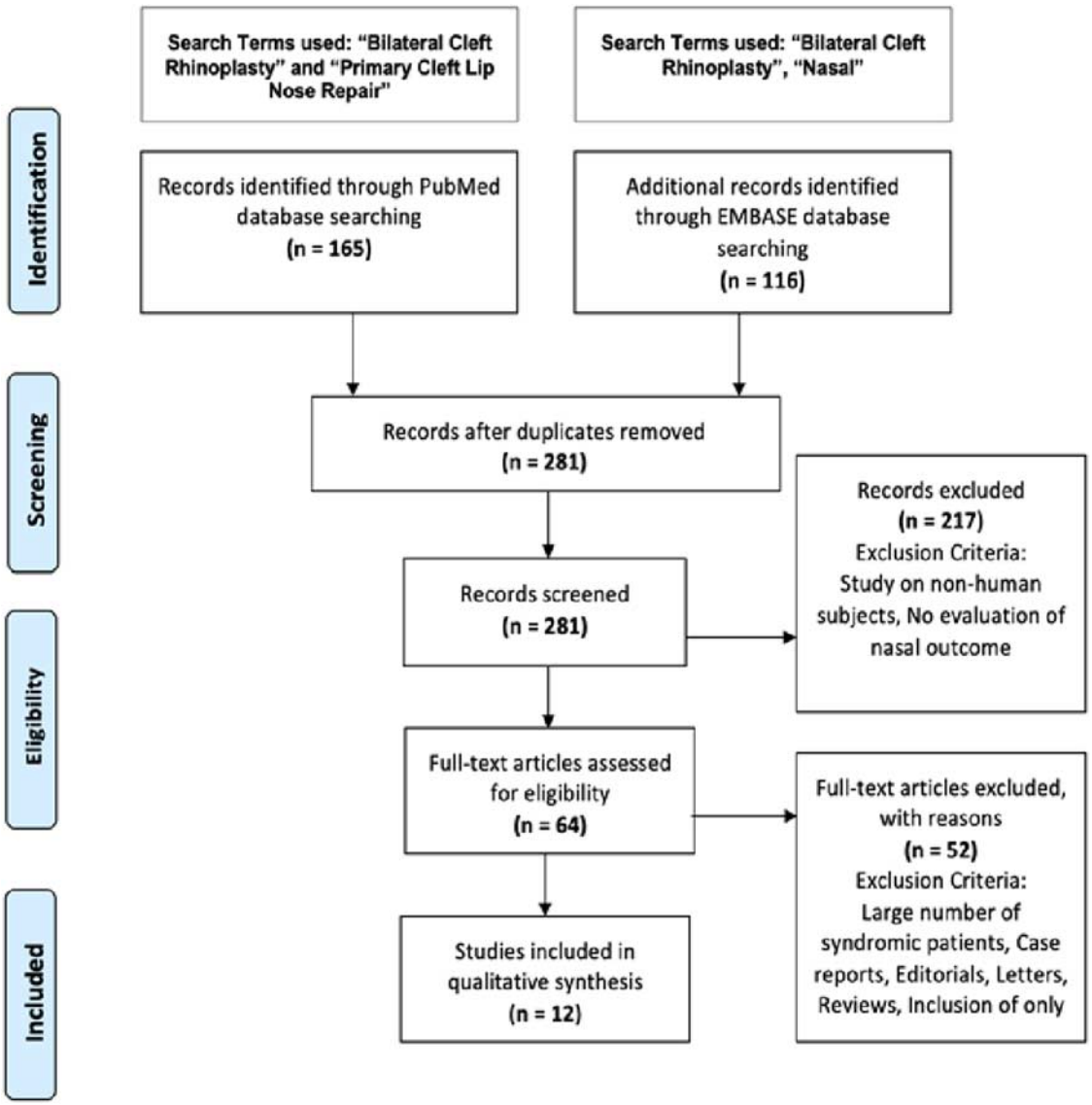


**FIGURE 1.** Preferred Reporting Items for Systematic Reviews And Meta-Analyses (PRISMA) flowchart of selected studies for systematic review.

were close to the mean for their age-matched counterparts, specifically for nasal protrusion, columellar length, and lip height.[19] Measurements taken directly using serial castings at 5 different points in time showed no statistical difference in experimental group relative to age-matched controls with respect to nasal tip protrusion, alar base width, alar width, columella length, and columella width.[18] Comparison of columella length over time in PR cleft patients with age-matched controls showed an increase, resulting in larger than normal nostril size and drift of the columellar base.[7]

## Nasal Development

Eight studies evaluated nasal growth and development overtime[7,13–19]. Follow-up measurement timing ranged between 2.5 years[15] to 19 years later[13,16].

Method of growth measurement included subjective author or team evaluation,[7,13–15] comparison of columellar lengths,[7] 2D indirect anthropometric measurements,[14,15,17] 3D indirect anthropometric measurements,[16,20] serial casting for measuring anthropometric distances,[18] and direct anthropometric measurements.[19] None of the 8 studies found restriction in nasal development over time.

## Need for Secondary Rhinoplasty

Four studies discussed the percentage of patients in which secondary rhinoplasty was not performed.[6,13,16,18] Only studies with a follow-up period of at least 6 years were included. The follow-up period ranged from 10 years[6] to 18 years.[13,16] The percentage of patients in whom the revision rhinoplasty was averted ranged from 23.10%[11] to 100%.[18]

# DISCUSSION

Although there has long been a concern surrounding PR, all 12 studies in our systematic review support performing PR in bilateral cleft patients; this support varied from eliminating need for secondary rhinoplasty, decreasing the size of secondary procedures required, and curbing long term psychosocial burden.

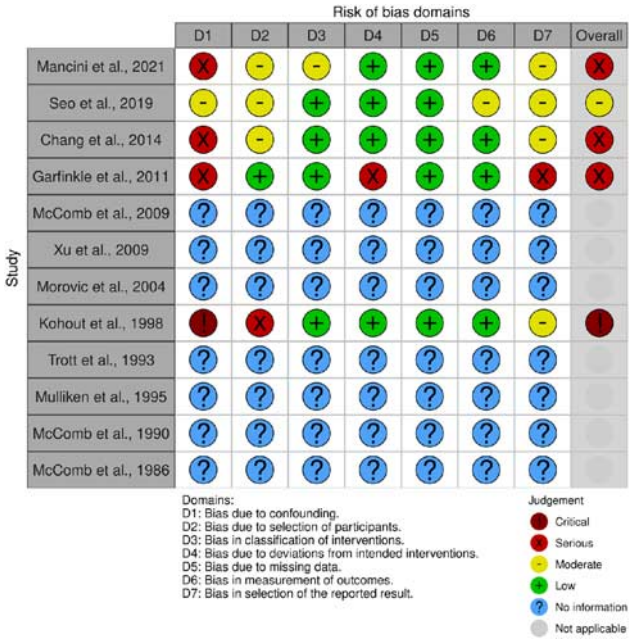


**FIGURE 2.** Traffic Light Diagram showing risk of bias in included studies.

However, there are some definite weaknesses among the reviewed literature. Studies ranged from years 1986 to 2019 with widely variable study designs, surgical techniques, and measurements taken; with only 2 of the studies implementing the use of technology for precise measurements. These differences made some articles difficult to compare directly. The main weakness was the volume of long-term outcomes—only 4 studies followed patients long enough to consider avoidance of secondary procedures. These limitations are further illustrated in the risk of bias determination. A majority of articles did not provide enough information for adequate risk of bias scoring, and those that did had either serious or critical risk of bias. On the basis of this alone, this review cannot make a high-quality recommendation, as a review is only as good as the studies it reviews. The main value of this review is look at trends in the available literature, highlighting the gaps in the evidence as it stands.

To minimize confounding and bias, measured objective outcomes are the most reproducible and comparable findings. Eight of the reviewed studies reported these, with a wide range of follow-up time; 5 articles followed patients to the age of 6 months to 3 years,[14,15,19,20] which is likely too short to make decisions about the long-term efficacy of PR. The remaining three articles evaluated patients aged 9 and older,[7,16,17] all showing positive long-term results.

The authors utilized widely variable anthropometric measurements. Three studies utilized conventional 2D photography,[14,15,17] which is difficult, as it requires standardization and can easily be affected by differences in user, lighting, and focus. Chang et al[14] acknowledged this error in photo variance by using correlation analysis of photos taken of the same patient one day apart. The newest studies used 3D imaging—the most useful method to take anthropometric measurements.[16,20] Future studies should aim to use this technology given its superiority over older techniques.

The ability to analyze nasal growth restriction as a consequence of PR requires follow-up of patient populations through puberty; 5 of the reviewed articles evaluated patients aged 12 to 19 with a total of 138 patients.[7,13,16–18] None of these showed interference in nasal growth, with 1 even noting an increase in nasal parameters overtime, specifically an increase in columellar length, compared with age-matched norms.[7] Another study that followed patients to maturity at 19 years old discovered that there were no significant differences in nasal surface area and volume when compared with normal subjects.[16] Garfinkle et al[18] followed patients to 12.5 years, finding parallel and proportional growth relative to the noncleft control group. Although our review identified only 5 studies that evaluated nasal growth objectively in BLCP patients, the general consensus reports no restriction. Although these results are promising, this is an area where large, prospective, long-term studies are warranted to truly identify the effects of PR on nasal growth. Surgeons performing the procedure should continue to follow post-operative outcomes with objective growth measurements to track developmental changes. Some studies noted changes to nasal morphology in the short term, many of these changes normalized overtime and into puberty.[17,18] It would be interesting to see whether these findings were reproducible in a larger, long-term cohort of patients; this certainly would change our counseling regarding what to expect after PR.

We doubt that the question of whether PR results in avoidance of secondary rhinoplasty will ever be definitively answered. This is because the decision to perform a secondary rhinoplasty is strongly affected by surgeon bias, patient preferences, and practice pattern variation. The available data are not adequate to determine whether the PR can truly be a final rhinoplasty. The majority of included studies that followed patients into maturity demonstrated avoidance of secondary rhinoplasty; 4 studies followed 158 patients to a mean age of 15 years, with 77% of patients avoiding secondary rhinoplasty.[6,13,16,18] The largest study[18] demonstrated that at 12.5 years of age, patients had near normal nasal appearance and none required secondary rhinoplasty. In a relatively short-term study in 1986,[6] it was found that only 1 patient out of 33, followed until the age of 10 years or older, required secondary rhinoplasty to adjust the columellar base. Seo et al[16] found that 76.9% of patients had an intermediate rhinoplasty around preschool age, whereas Kohout et al[17] followed patients to about 15 years of age, but did not comment on secondary procedures. The broad disparity in these results indicates that further work is needed; but given the limitations of patient and surgeon preferences, we will unlikely ever be able to identify a technique that would truly avoid secondary rhinoplasty in most patients.

"Primary rhinoplasty" is not the same for all surgeons—a variety of procedures were used in the reviewed studies, none of which included septoplasty. The main weakness in this type of study methodology is that we cannot control for the type of intervention. However, even more concerning is that studies did not compare PR with patients with a cleft who did not undergo PR. However, some logical assumptions can still be made. Many studies were able to demonstrate similarities in many aspects with PR and normal controls. This would seem to be an improvement over patients who had no rhinoplasty, whose noses are often very different than those of children without a cleft. Thus, it logically follows that PR does have a positive effect on nasal appearance. However, without a cohort study comparing PR with no PR, we are unlikely to have definitive answers to what exactly is affected by PR and how much.

The minimal comparative literature available does not currently indicate an ideal technique at this time. Kohout et al[17] found a significant difference between Mulliken and Trott techniques, with improved columellar length and nasal projection in Mulliken's technique. Chang et al[14] identified that progressively more aggressive techniques resulted in improved outcomes, with NAM plus PR with overcorrection resulting in a nasal appearance most similar to their control group of cleft

patients who had undergone palatoplasty only. Future work is required to identify the ideal PR technique, using objective measurements and comparative data from multiple centers.

## CONCLUSION

Literature regarding PR in bilateral cleft lip is more limited than that of unilateral cleft lip. Most available studies support the practice of performing a PR and this does not seem to restrict nasal growth. However, there are serious limitations to available literature amount and quality, highlighting a need for larger comparative studies to answer more specific questions regarding the practice of PR, including optimal technique or avoidance of secondary revision rhinoplasty.

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