

Differences in the clinical appearances of white versus black patients with facial cleft deformities: A retrospective study of a South African clinic

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ABSTRACT

Objective: The purpose of this study was to determine the difference in prevalence of the different categories of facial cleft deformities between the white and black patients in a database of 2806 cleft cases at the University of Pretoria. No variation of clefts between these two groups has ever been compared previously. For this purpose, the clefts were classified according to the system described by Bütow in 1985.³

Material and Methods: Records of 2806 patients attending the university's cleft lip and palate clinic, between August 1983 and February 2006, were reviewed. The study group included cleft patients with syndromic and nonsyndromic CL, CLA, CLAP, hP, hPsP, sP, COMBI clefts with or without oblique or transverse facial (or Tessier) clefts. Very few of the cases presented with speech problems only, but with no clefts. The cleft categories of cleft lip, cleft palate and cleft lip and palate, as well as their subdivisions, were analysed.

Results: Of these cleft patients, 2003 were white patients and 665 were black patients; the rest were Indian, Coloured and East-Asian patients. In the black and the white population groups (n = 2668), there were more males with cleft in the white group (58,2%), but more females with clefts in the black group (54,9%). The most common cleft type generally was the cleft lip, alveolus and palate cleft (CLAP) with a 434% prevalence of white patients and 296% of black patients. The most common cleft in the black patients was the cleft palate at 435%, which was only recorded in 35,0% of the white patients. The frequency of the other orofacial clefts in decreasing order was: sP 19,4% for white and 21,2% for black; hPsP 15,2% for white and 21,2% for black; CLA 9,7% for white and 19,8% for black; CL 5,9% for white and 4,5% for black and combinations of different orofacial clefts (COMBI), 6,0% for white and 2,6% for black. The isolated hard palate cleft (hP) occurred very rarely (0,4% for white and 1,1% for black). The left side of the face was more often afflicted (left to right 51,6% to 28,5% for white; and 35,0% to 37,9% for black). In 0,5% (white) and 3,9% (black) median clefts were observed.

Conclusion: A retrospective study was done of 2806 facial cleft cases in order to analyse and then determine the frequency of specific clinical appearances in the white and the

black patients (2668 cases). There was a considerable variation between the groups of orofacial clefts occurring in the two groups.

Key words: cleft variation, white black cases, categories, cleft lip, cleft lip and palate, cleft lip and alveolus, cleft palate, oblique facial clefts

INTRODUCTION

Cleft lip and palate or facial cleft deformities are among the most common congenital malformations and were already recognised in ancient times. Since these orofacial and oblique facial clefts are easily recognised and described, they have become one of the most intensively studied congenital malformations worldwide.¹

The clefts found in the white population included in the studied database, should conform to the general worldwide trend. However, the cleft deformities of the black population may differ from and may have more variations than the better-known trends and no literature of this nature has previously been published. The following population groups will be referred to in this publication: Asian, 2.5% of the population - mainly people of Indian descent; Blacks, 79% of the population - descendants of African peoples who migrated in a southerly direction from central Africa; Coloureds, 8.9% of the population - people of mixed parentage, mainly descendants of the indigenous Khoikhoi people, Malayan slaves and the White settlers; and Whites, 9.5% of population - descendants of the European settlers, mainly Dutch, British, German, French Portuguese, etc.²

The aim of this study was a retrospective analysis and determination of incidence of various categories of clefts of 2668 black and white patients, who were found in the total database of 2806 cleft patients registered at the University of Pretoria, over a period of almost 23 years (1983 to 2006). The cases were classified according to the clinical classification system described by Bütow in 1985 and 1995^{3,4} (Table 1).

Type of Cleft	Abbreviation	Subdivision
Cleft lip	CL	- uni-, bilateral (uCL, bCL) - left, right (l-CL, r-CL) - partial, total - median - lateral
Cleft alveolus	CA	- uni-, bilateral (uCA, bCA) - left, right (l-CA, r-CA) - partial, total - median
Cleft lip and alveolus	CLA	- uni-, bilateral (uCLA, bCLA) - left, right (l-CLA, r-CLA) - partial, total
Cleft lip alveolus and palate	CLAP	- uni-, bilateral (uCLAP, bCLAP) - left, right (l-CLAP, r-CLAP) - partial, total
Isolated hard palate	hP	
Hard and soft palate	hPsP	- left, right, bilateral
Soft palate	sP	- total sP cleft - partial sP and/or bifid uvula - submucosal
Combinations of different orofacial clefts	COMBI	Any combination of above-mentioned, as well as other cleft deformities which cannot be placed in above-mentioned categories
Oblique or transverse facial clefts (Tessier 4)	Oblique facial	
Speech problems or velopharyngeal insufficiency	NC	(no clefts - cases with speech problems)

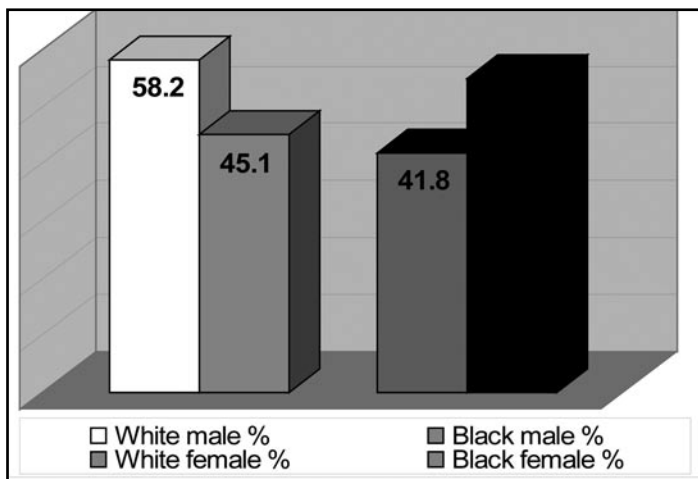


Figure 1: Distribution of the different sex and population groups Ninety-five (3,6%) patients were born with combined orofacial and oblique facial, median or lateral (transverse) facial clefts (0,8% in white, 4,5% in black)

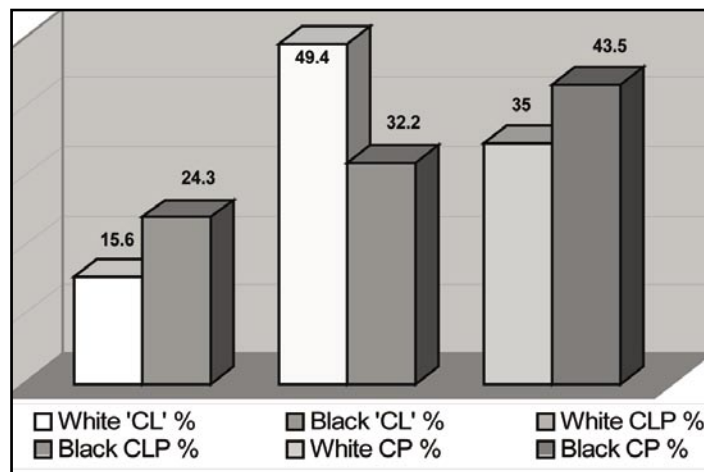


Figure 2: Frequency of facial cleft categories according to the main categories of the ICD-DA (1995) for white and black cleft patients

PATIENTS AND METHODS

Since the establishment of the Cleft Lip and Palate or Facial Cleft Deformity Clinic at the Department of Maxillo-Facial and Oral Surgery of the University of Pretoria in 1983, up until February 2006, 2806 patients presented for treatment of their facial clefts. It is estimated that the majority of the patients afflicted with cleft deformities in South Africa have attended this multi-disciplinary clinic. Apart from the general demographic information (age, population groups, gender, etc.) the following data are also recorded during contact with

the patient: pregnancy history of the mother, exposure by the mother to possible etiological agents, genetic history, associated malformations, clinical appearance and classification of the cleft according to the classification system of Bütow (1985),³ treatment the patients have received, and complications. This data was captured and retrospectively analysed using Microsoft Excel and Access computer programmes. The database for this study consists of 2668 black and white patients. Most of the patients attending the clinic are either referred by healthcare professionals from surrounding state and private hospitals, or are sent by general practitioners and various other specialties from the private health sector. Some

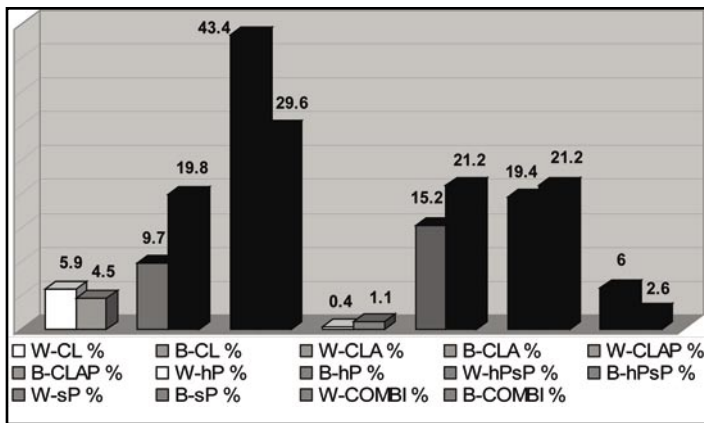


Figure 3: Frequency of facial clefts in patients categorised according to Bütow

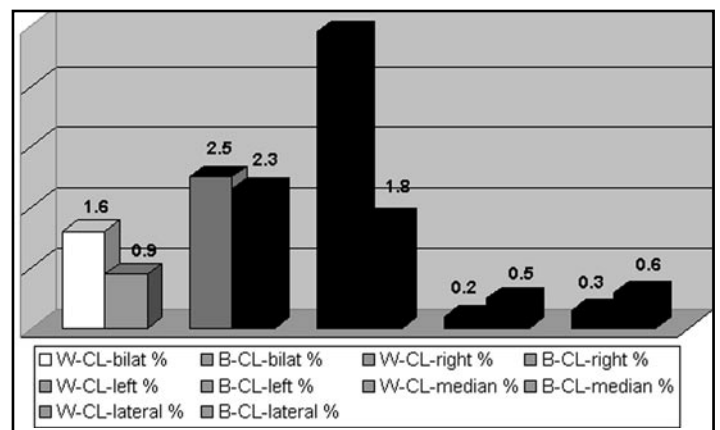


Figure 4: Distribution of the subgroups of cleft lip (CL) in the database

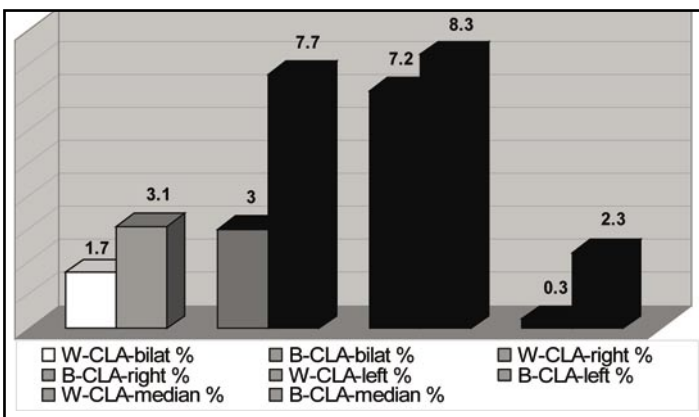


Figure 5: Distribution of the different subgroups of cleft lip and alveolus (CLA) in the database

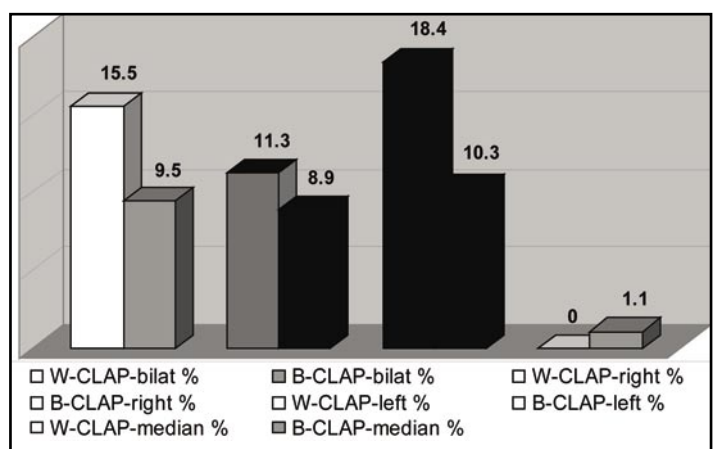


Figure 6: Distribution of the different subgroups of cleft lip, alveolus and palate (CLAP) in the database

of these patients are self-referrals. All of them are evaluated, treated (if consented to by the patient or parents) and followed-up, preferably annually.

RESULTS

Of the 2668 black (665 = 24.9%) and white (2003 = 75.1%) patients who attended the Cleft Lip and Palate or Facial Cleft Deformity Clinic, all were diagnosed at the initial registration, with orofacial and/or oblique facial clefts (including transverse facial or Tessier's cleft⁵) or speech problems involving the soft palate. The population group versus sex distribution has been recorded in Fig. 1.

Fig.2 shows the frequency of traditional facial cleft categories according to the categories of the ICD-DA (1995)⁶ [cleft lip, cleft lip and palate and cleft palate], whereas Fig.3 highlights the clinical categories according to Bütow (1985)³ (Table 1) for facial cleft deformities. The results, as indicated in Fig. 3, have been divided to show the different types of cleft deformities based on the clinical classification. The most common types were: cleft lip, alveolus and palate (CLAP), followed by cleft soft palate (sP), cleft hard and soft palate (hPsP), cleft lip and alveolus (CLA) and cleft lip only (CL). Combination clefts (COMBI), oblique facial clefts and isolated cleft of the hard palate (hP) occurred the least in the sample. Fig.4 presents the distribution of the different subgroups [excluding man-

dibular cleft lip] of cleft lip (CL) in the database, (147 cases, an overall incidence of 5,5% of the database for each of the black and white population groups), namely bilateral cleft lip (CL-bilat), unilateral right-sided cleft lip (CL-right), unilateral left-sided cleft (CL-left), and median cleft lip (CL-median). The transverse facial or lateral cleft lip (lateral facial or Tessier⁷ cleft⁵) was found in 0,3% (white) and 0,6% (black) patients. A single black patient presented with a bilateral transverse facial cleft lip and another black patient with a median cleft lower (mandibular) lip.

The overall incidence rate of cleft lip and alveolus (CLA) was recorded 334 in patients. This cleft type is traditionally placed in the cleft lip category⁶. However, a dento-alveolar cleft is present and in most cases an anterior nasal floor reconstruction is required as part of the primary cleft reconstruction. A bone graft (therefore, an early or late secondary osteoplasty) is the next stage to create a continuous dento-alveolar arch in the child or adolescent (part of the secondary or tertiary cleft reconstruction). The different subdivisions of a cleft lip and alveolus (CLA) are: bilateral cleft lip (CLA-bilat), unilateral right-sided cleft lip (CLA-right), unilateral left-sided cleft (CLA-left) and median cleft lip (CLA-median) (Fig.5).

The most frequent type of orofacial cleft was the cleft lip, alveolus and palate (CLAP) with a prevalence of 1098 cases

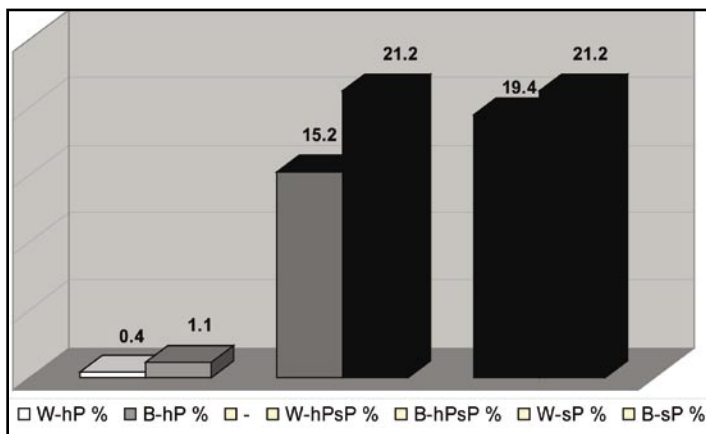


Figure 7: Distribution of the different subgroups of cleft palate (hP, hPsP, sP) in the database



Figure 8: Typical appearances of an isolated hard palate cleft (hP)

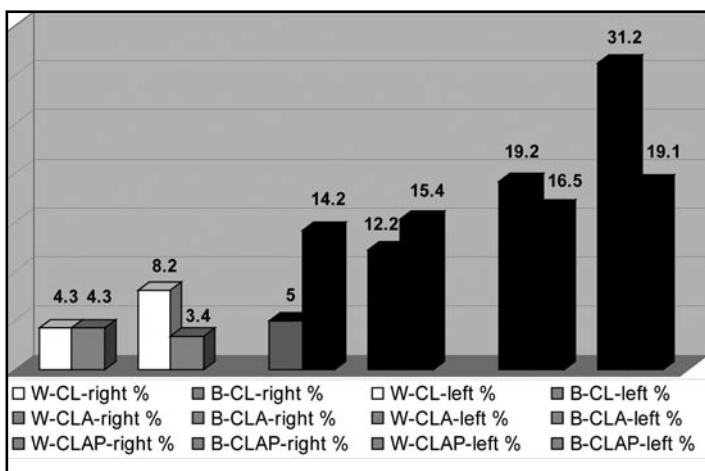


Figure 9: Distribution of the different types of UNILATERAL clefts (CL, CLA, CLAP) (right and left side) in the database

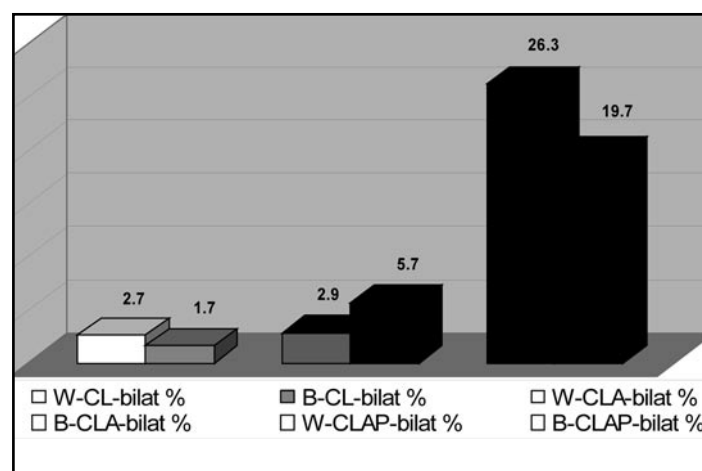


Figure 10: Distribution of the different types of BILATERAL clefts (CL, CLA, CLAP) in the database.

(Fig.6). The cleft lip, alveolus and palate are subdivided into the following subgroups: bilateral cleft (CLAP-bilat), unilateral right-sided cleft (CLAP-right), unilateral left-sided cleft (CLAP-left) and median cleft (CLAP-median). The abbreviation COMBI represents combinations of cleft types occurring in one person. Combinations of different cleft types (COMBI) (for example: CL + sP, without an hP; unilateral CLA + bilateral hP + sP; CL + partial A + partial hP + sP; bilateral CL + unilateral A + hPsP, etc) occurred in 117 white and 17 black patients respectively, of the studied database (Fig.3).

The palatal clefts (hP, hPsP and sP) (966 cases or 36.2% of the total database for black [283] and white [683] patients), may be subdivided into the following main groups: very rare isolated hard palate cleft (hP), hard combined with soft palate cleft (hPsP) and soft palate cleft (Fig. 7). With an overall prevalence of 0,5% (14 cases, equal in both population groups) the isolated cleft of the hard palate (hP) (Fig. 8) was one of the rarest orofacial cleft types. The soft palate cleft (sP) occurred in 544 patients or 20,4% of the total number of cases. Soft palate (sP) clefts may either be total (full length cleft, 84,4% - white; 81,4% - black) or partial (in its length) - the latter includes the bifid uvula. Together, the total and partial sP had the highest prevalence of sP clefts. The subtype submucosal soft palate cleft (mucosal fusion,

but no muscle connection, in the aponeurosis) was recorded in 27,5 % (white) and 14,8% (black) of the sP cases and may occur in combination with a partial sP cleft or on its own where only the muscle is involved without any mucosal involvement. Unilateral clefts, right or left side, were recorded in 78,4% (1179) of CL, CLA and CLAP. Amongst these clefts, the uCLAP was the most common (50,4% - white; 35,6% - black) - highest: CLAP-left – 31.2% - white).

The uCLA was recorded in 17,2% (white) and 29,6% (black patients). The left unilateral cleft (CLA-left) was the most common of these clefts, and was found in 15,4% of the black patients. With a prevalence of only 12,5% (white) and 7,7% (black) for the uCL (lowest recorded for the l-CL – 3,4% - in black patients), this type of unilateral cleft had the lowest incidence (Fig. 9). One of the rarest unilateral cleft types is the lateral or transverse uCL (Tessier⁷ cleft⁵) (0,3% - white; 0,6% - black). The ratio between the right side versus the left side cleft lip, as in uCL, uCLA and uCLAP, is 35,5% versus 64,5% in white patients and 48,0% versus 52,0% in black patients. Bilateral clefts were recorded in 30,7% (462) of all the CL, CLA and CLAP. Amongst these clefts the bCLAP was the most common type. Bilateral CLA accounted for only 2,9% in whites. However, it occurred in 5,7% of the black patients. In contrast, the bCL was recorded in only

1,7% of the black patients (Fig.10). The rarest bilateral cleft was found as a transverse facial bCL (lateral facial or Tessier⁷ cleft⁵) - one case, 0,5% of cleft lips or 0,04% of the studied cleft database. Only 34 (2,9%) of the 1179 CL, CLA and CLAP patients presented with median clefts. [CL (0,3% - white; 0,3% - black) and CLA (only 0,4% for white patients; however, 1,3% for black patients)]. Six median CLAP, were recorded in black patients. The rarest median cleft lip was that of the mandibular lip, found in one black patient, or 0,5% of CL or 0,04% of the total database.

DISCUSSION

Clinicians' common goal is a better understanding of this human malformation in order to provide a comprehensive treatment. The traditional classification of facial cleft deformities is derived from the developmental, embryological or morphological perspective^{8,9} and is divided into clefts of the primary palate (which is the lip and the dento-alveolar arch) and secondary palate (which includes the hard palate and soft palate).^{10,11} They are therefore categorised into cleft lip, cleft palate and the combination of cleft lip and palate. A developmental or embryological classification is therefore not entirely suitable for this purpose, since cleft lips, either with or without cleft alveoli, require different treatment modalities and therefore more detailed classifications are necessary. This situation also applies to the cleft palate as it is essential that hard and soft palate clefts, as well as unilateral and bilateral cleft hard palate, are categorised separately. Each category of clefts requires individual clinical and surgical treatments to ensure optimal reconstruction of these facial cleft deformities. For this purpose, all the cleft cases in this database were evaluated according to a detailed clinical,^{3,4} and not a developmental, classification system.⁶

Sixty-one percent of all the patients (2806) attending this Facial Cleft Clinic are within the age-range 0 to 1 years. This underlines the demand for primary treatment of infants registered at the clinic. Older children were referred for diagnostic reasons or for the exclusion of previously misdiagnosed or missed sP clefts. Other older cleft-afflicted individuals, especially adolescents, were referred for secondary cleft reconstruction after they had received their primary cleft repair elsewhere in the country or in neighbouring states. This retrospective analysis of cases registered with the Facial Cleft Clinic differentiates the various cleft categories found in 2668 black and white cleft patients.

Variations of orofacial clefts

The cleft lip, alveolus and palate (CLAP) is the most frequent cleft type (43,4% - white; 29,6% - black). A study in British Columbia revealed similar results (40,6%),¹² while a Japanese publication indicated a higher frequency of 47,3%¹³ as did a Mexican publication,¹¹ where this cleft type was registered in 54,8% of cases. The Chinese Shanghai population had an incidence of 65%,¹⁴ which is considerably higher than the occurrence rate observed in this cleft database for the white patients. In contrast, the frequency of these clefts in the Pakistani population was much less at 34%.¹⁵ This is

more compatible with the incidence of 29,6% for the black patients in this database, who had the lowest recorded figure in all these studies.

The second most frequent cleft type was the soft palate cleft (sP) (19,4% - white; and 21,2% - black). The combined hard and soft palate cleft (hPsP) type (15,2% - white; 21,2% - black) presented with a slightly lower prevalence than the soft palate cleft (sP) for the white patients. The rare isolated hard palate cleft (hP) was recorded at 0,4% (white) and 1,1% (black). Therefore all palatal clefts (hP, hPsP, sP) combined, for both the white and the black patients, had a high prevalence of 39,3% (35,0% - white; 43,5% - black). Again the study done in British Columbia revealed similar results for white patients (36,5%),¹² whereas the following studies presented with quite different results: Iran 17,4%;¹⁸ Japan 18,7%;¹³ Mexico 20,2%;¹⁰ Pakistan 24%;¹⁵ Denmark 25%;⁸ and Australia (Victoria) 35.5%.¹⁶ The black patients in our study therefore presented with a much higher prevalence of palatal clefts. Due to the fact that the soft palate (sP) cleft may vary considerably in its cleft length, this cleft type was divided into three subtypes. The total-type of soft palate clefts (sP) was observed in 84,4% (white) and 81,4% (black) cases.

The shortest partial sP cleft type was the bifid uvula. A sub-mucosal sP was found in 27,5% (white) and 14,8% (black) patients, with or without a partial cleft. The combination of submucosal and partial sP clefts, including the bifid uvula, was recorded less often. It could be that these subtypes of sP clefts are more common in the general population, but due to their low impact as far as speech impairment or feeding problems is concerned, these cleft patients most probably do not seek medical attention as much as other cleft patients. The average incidence of the cleft lip and alveolus (CLA) was 9,7% (white) and 18,8% (black). This is a considerable variation between the white and black patients in this database. In discordance to Iregbulem,¹⁷ who reported a CL incidence of more than 40% in his/her analysed Nigerian population, an incidence rate of only 5,9%/4,5% (white/black) CL was found within this cleft database. It is possible that this particular author,¹⁷ as well as possibly other authors (British Columbia 22,9%;¹² Mexico 25,0%;¹⁰ Australia (Victoria) 27,3%;¹⁷ Japan 34%;¹² Iran 34,9%;¹⁸ Pakistan 42%¹⁵) categorised their cleft cases only according to the developmental (embryological) classification. This might account for a higher occurrence rate, since their CL and CLA cases were most probably grouped together. If the database of this study were to be adjusted accordingly, there would be 15,6% (white) and 24,3% (black) (CL + CLA) cases, which would fall within the figures of the British Columbia study.¹² Eleven patients presented with a rare lateral or transverse CL, therefore an overall incidence of 0,4%. A median cleft type was detected in 34 patients (1,3% overall incidence rate: 0,4% - white and 0,9% - black). The ratio between the right side versus the left side cleft lip, such as in uCL, uCLA and uCLAP is 35,5% versus 64,5% in white patients (which is nearer to the one-third versus two-third ratio), and 48,0% versus 52,0% in black patients (which is nearer to the

half- versus-half ratio). Some of the rarest cleft types were recorded, such as the isolated hard palate cleft (hP), with an overall incidence of only 0,4% (white) and 1,1% (black); a bilateral lateral (transverse) facial cleft lip (CL) (0,04% - in a single black female), a median mandibular cleft lip (CL) (0,04% - in a single black female), and a hyobranchial cleft (cleft between the hyoid bone and suprahyoid muscles - 0,04% - in one white female). Apparently only 68 cases of median mandibular or lower cleft lip have been reported in the literature.¹⁹

Oblique facial clefts

Forty-seven (1,8%) patients found in this database had oblique facial clefts, with or without orofacial clefts.

CONCLUSION

Orofacial and/or oblique facial clefts were recorded in all the black and in all the white patients extracted from a total database of 2806 cases. More cleft deformities were recorded in white males (58,2%), while more black females (54.6%) were recorded with cleft deformities. The unilateral clefts (CL, CLA, and CLAP), right versus left, had a ratio of nearly one-third: two-thirds for the white patients, and nearly the half: half ratio for black patients. The most common cleft type was the CLAP in white patients; with, however, significant differences between these two race groups. After combining the different cleft palate deformities (hP, hPsP, sP), the black patients presented with far more cleft palate deformities than white patients. Whereas the white cleft patients had the highest incidence of CLAP, the black patients had a very high incidence of CLA.

Generally, the more rare cleft deformity types were seen in black patients, such as the median cleft, one mandibular median CL, unilateral transverse CL and one bilateral transverse cleft lip. The only hyobranchial cleft was registered in a white female. This study reveals a distinctive difference in the prevalence of types of cleft deformities between these two population groups.

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CONFLICT OF INTEREST. NO CONFLICT OF INTEREST WAS DECLARED BY THE AUTHORS

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