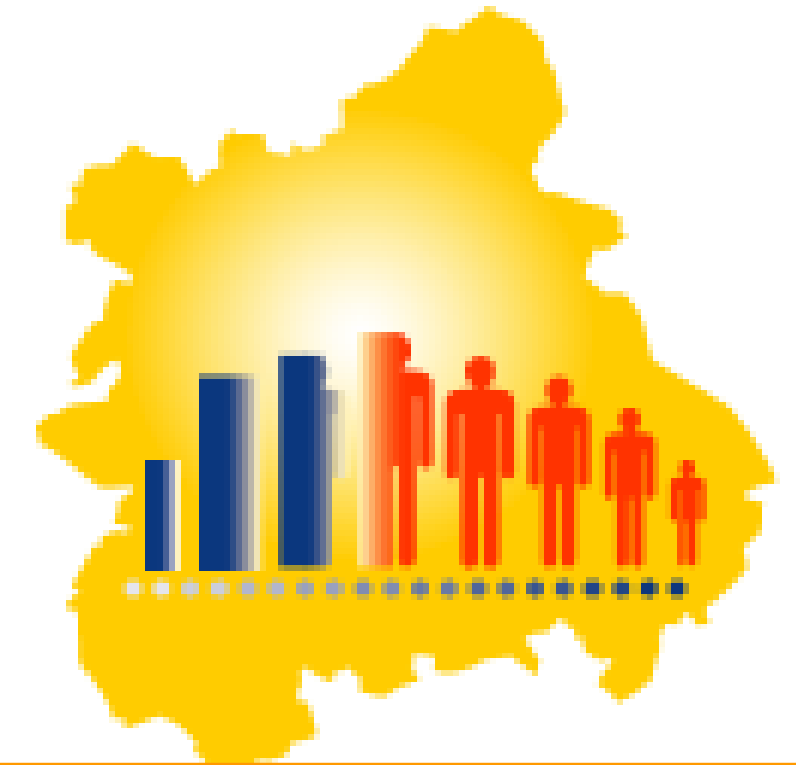# Incidence of Sarcomas of the Facial Skeleton

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ABSTRACT

There are no national statistics of the incidence of rare malignant bone sarcomas of the facial skeleton. Although cancer registries record all cancers diagnosed in England, limitations in ICD-10 coding mean that sarcomas of the face cannot be distinguished from sarcomas of the back of the skull. Such cases are coded as *malignant neoplasm of bones of skull and face* (C41.0) and *malignant neoplasm of mandible* (C41.1) using ICD-10. Cancer registry data and Hospital Episode Statistics (HES) data allow an estimate of the incidence of these tumours by distinguishing facial tumours coded to C41.0 from those of the back of the skull.

METHOD

The number of facial C41.0 cases can be estimated by extrapolating the known proportion of facial sarcomas in a studied cohort to the total number of C41.0 cases registered in England. Proportions were derived for each morphology grouping using two cohorts:

Figure 1: Bones of the human skull, demonstrating those classed as facial
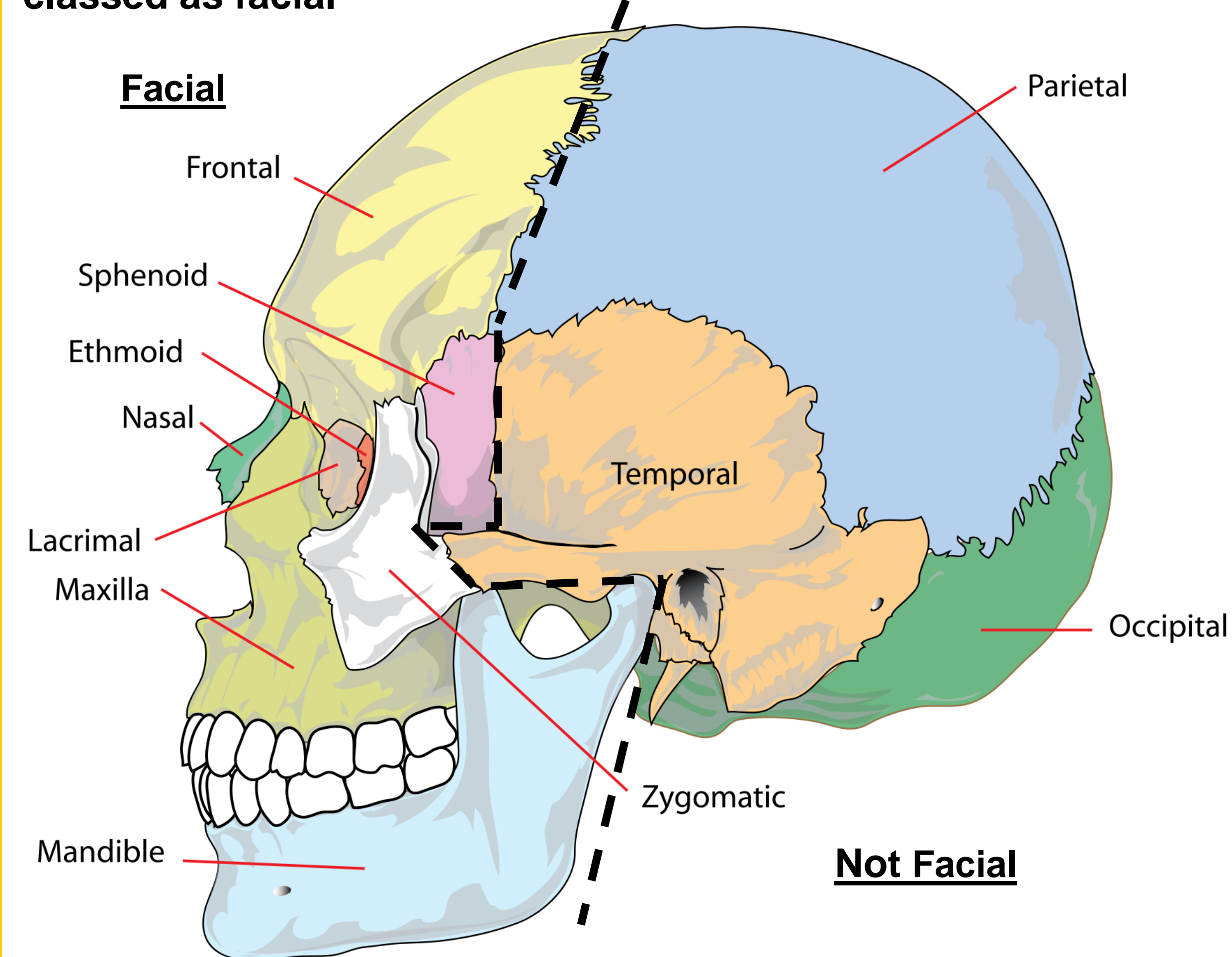
West Midlands
- Cohort of 75 patients diagnosed with C41.0 between 1979 and 2009
- Pathology reports within the West Midlands cancer registration database examined for details of tumour location
  ➡ Categorised as 'Facial', 'Not Facial' or 'Unknown'

HES
- Cohort of 212 patients diagnosed with C41.0 between 2000 and 2007
- Operational information from HES records was examined for key words relating to the face (e.g. 'face', 'maxilla', 'nasal') or skull ('cranium', 'temporal bone')
  ➡ Categorised as 'Facial', 'Not Facial' or 'Unknown'

The total number of facial sarcomas was derived by combining all C41.1 cases and the estimated number of facial C41.0 cases. The estimates were calculated by applying the derived proportions of facial sarcomas to all C41.0 cases within each morphology grouping. 'Unknown' cases were treated as 'Not Facial' when extrapolating the proportions within each morphology group.

Table 1: Grouping of ICD-O-3 codes

Morphology Grouping	ICD-O codes				
Ameloblastoma	9310/3				
Carcinoma	8010/2	8010/3			
Chondrosarcomas	9220/3	9221/3	9230/3	9231/3	9240/3
Chordoma	9370/3				
Ewing Sarcoma	9260/3				
Fibrous Sarcomas	8810/3	8811/3	8830/3	8840/3	
Neoplasm malignant	8000/3	8000/9			
Osteosarcomas	9180/3	9181/3	9182/3	9184/3	9190/3
Other	8002/3	8003/3	8070/3	8140/3	8246/3
	8525/3	8800/3	8801/3	8890/3	8900/3
	8910/3	8920/3	8982/3	9040/3	9064/3
	9120/3	9133/3	9250/3	9270/3	9290/3
	9364/3	9400/3	9470/3	9500/3	9561/3

RESULTS

Table 2: Morphology types of West Midland C41.0 tumours and the ensuing proportion classified as 'facial'

ICD-O Group	C41.0 sub-catagories:			Total	% Facial
	Facial	Not Facial	Unknown		
Ameloblastoma	2	0	0	2	100%
Carcinoma	0	1	0	1	0%
Chondrosarcomas	5	20	2	27	19%
Chordoma	0	0	13	13	0%
Ewing Sarcoma	0	1	1	2	0%
Fibrous Sarcomas	1	0	0	1	100%
Neoplasm malignant	0	2	0	2	0%
Osteosarcomas	15	6	1	22	68%
Other	3	2	0	5	60%
Total	**26**	**32**	**17**	**75**	**35%**

Table 3: Morphology types of HES C41.0 tumours and the ensuing proportion classified as 'facial'

ICD-O Group	C41.0 sub-catagories:			Total	% Facial
	Facial	Not Facial	Unknown		
Ameloblastoma	6	0	1	7	86%
Carcinoma	3	1	0	4	75%
Chondrosarcomas	20	28	15	63	32%
Chordoma	6	24	7	37	16%
Ewing Sarcoma	2	4	7	13	15%
Fibrous Sarcomas	3	1	0	4	75%
Neoplasm malignant	9	4	8	21	43%
Osteosarcomas	22	10	5	37	59%
Other	15	5	6	26	58%
Total	**86**	**77**	**49**	**212**	**41%**

The proportion of C41.0 sarcomas which could not be classified into sub-sites was consistent in both methods at 23%.

The number of cases in each morphology group is very small as these are rare tumours. This leads to uncertainty in the estimates.

The derived proportions of facial C41.0 cases from each method is shown in **tables 2 and 3**.

Figure 2 shows the estimated overall incidence of facial bone sarcomas. The average over the 18 year period is:

25 cases annually
[24 cases using the WMCIU data, 26 using HES, Not statistically significantly different]

The crude rate is

0.5 cases per 1,000,000 population
[0.49 cases using the WMCIU data, 0.54 using HES, Not statistically significantly different]

Figure 2: Estimated national occurrence of malignant facial sarcomas

CONCLUSION

Limitations in international coding systems mean sarcomas of the facial skeleton cannot be directly identified from cancer registry data. However, estimates of these rare tumours can be derived from national cancer registry data using local audits or HES data based on the proportion of facial cases for given morphology groups. Both data sources produce similar national estimates of approximately 25 cases of malignant facial sarcoma per annum. These predictions cannot be validated until coding systems are able to further reflect sub-sites.

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