A Clinical Audit on Compliance with British Society of Haematology Guidelines For Diagnosis And Management Of Philadelphia Negative Myeloproliferative Disorders

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INTRODUCTION

Myeloproliferative Neoplasms (MPN) are a group of disorders that affect multipotent progenitor cells

WHO classifies MPN based on the presence or absence of the BCR-ABL fusion gene on the Philadelphia chromosome

Philadelphia-negative MPN include:

- Myelofibrosis (primary, post-PV, post-ET)
- Polycythemia Vera
- Essential Thrombocythemia

OBJECTIVES

Primary objective:

To assess compliance with British society of haematology guidelines for diagnosis and management of Philadelphia negative myeloproliferative disorders

Secondary objectives include the description of:

Demographics and clinical characteristics of patients at time of diagnosis.

Type of management strategy

METHODS

The audit was conducted in the Clinical Haematology Department, New cross Hospital and Cannock chase hospitals Royal Wolverhampton NHS trust.

Record of 145 patients with a diagnosis of MPN (ET, PRV, PMF) was audited

Data Collection Period: August- November 2019

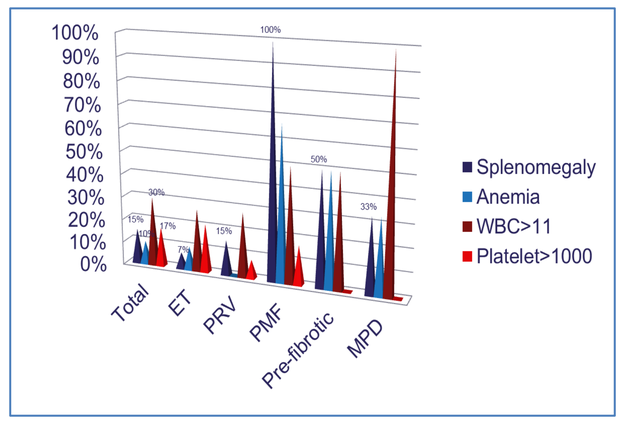
Data on patient demographics, clinical characteristics, management strategies and outcomes were collected from the clinical web portal.

RESULTS

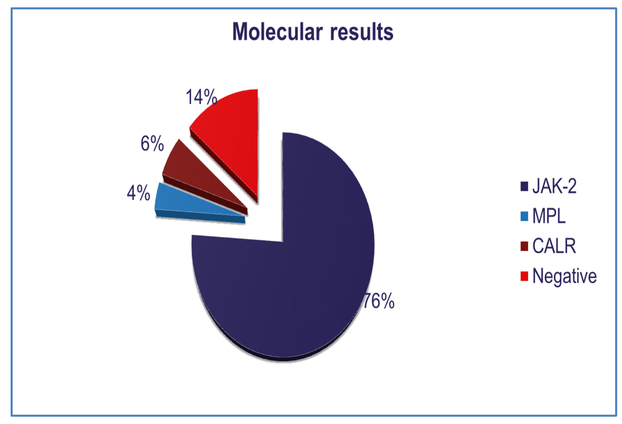
Characteristics	N=145
Male, n(%)	53 (37)
Female, n(%)	92 (63)
Median Age (years)	67
Age range (years)	34-93
Diagnosis *Essential Thrombocythemia (ET), n(%)* *Polycythemia Rubra Vera (PRV), n(%)* *Primary Myelofibrosis (PMF), n(%)* *Pre-fibrotic Myelofibrosis , n(%)* *Myeloproliferative disorder (MPD), n(%)*	 94 (65) 40 (28) 6 (4) 2 (1) 3 (2)

RESULTS

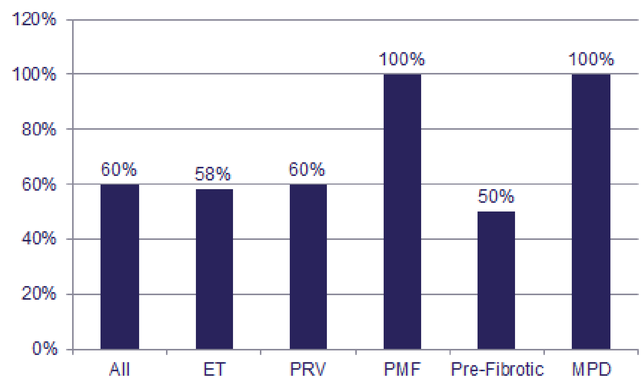
Symptoms	ALL (N=145) N(%)	ET (n=94) N(%)	PRV (n=40) N(%)	PMF (n=6) N(%)	Pre-Fibrotic MF (n=2) N(%)	MPD (n=3) N(%)
Asymptomatic	59 (41)	44(47)	11 (27)	1 (17)	1 (50)	2 (67)
Fatigue	16 (12)	12 (13)	3 (8)	1 (17)		
Headache /blurring of vision	10 (7)	4 (4.2)	6 (15)			
Dizziness	4 (3)	2 (2)	2 (5)			
Erythromelalgia	3 (2)	2 (2)	1 (2.5)			
Acquagenic pruritis/pruritis	8 (5.5)	3 (3.2)	5 (12.5)			
Body aches	5 (3)	4 (4.2)	1 (2.5)			
TIA/Stroke	3 (2)	3 (3.2)	0			
Parasthesia	3 (3)	2 (2)	1 (2.5)			
Shortness of breath	4 (3)	1 (1.1)		1 (17)	1 (50)	1 (33)
Unintentional Weight Loss	2 (1.3)	0	0	2 (32)		
PE	1 (0.7)	1 (1.1)	0			
Missing Records	22 (15)	14 (15)	8 (20)			
Not Documented	5 (3)	2 (2)	2 (5)	1 (17)		



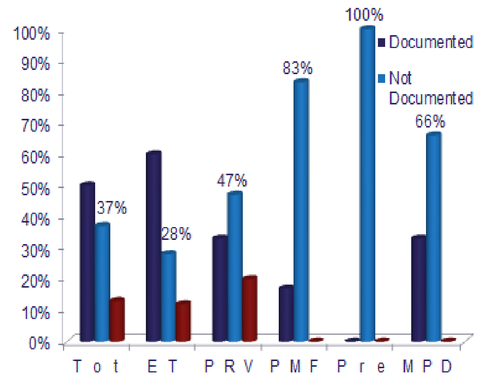
Work-up Done	All	ET	PRV	PMF	Pre-fibrotic MF (n=2)	MPD (n=3)
Bone marrow	96/141 (68%)	69/92 (75)	17/38 (42.5)	6/6 (100)	2 (100)	2 (66.6)
Acute phase Reactant	130/135 (96)	88/89 (99)	34/36 (94)	3/5 (60)	2 (100)	3 (100)
Iron Studies	130/135 (96)	88/89 (99)	34/36 (94)	3/5 (60)	2 (100)	3 (100)
Molecular	141/145 (97.2)	91/94 (97)	39/40 (97.5)	6/6 (100)	2 (100)	3 (100)



MDT Discussion



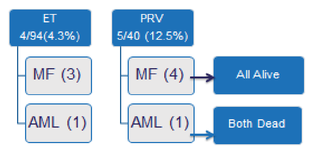
Risk Stratification



Management Strategies

Management Strategies	N (%)
Wait and Watch	22 (12)
Hydroxycarbamide	111 (61)
Anagrelide	5 (3)
Ruxolitinib	8 (4)
Busulphan	5 (3)
Radiophosphorus	1(1)
Venesection+Hydroxycarbamide	30 (16)

Disease Transformation



CONCLUSION

Compliance was satisfactory in:

- Diagnostic workup
- Treatment stratification for cytoreduction in high risk patients

Compliance was found to be poor in following areas:

- Doing bone marrow biopsy as part of workup
- Recording risk stratification
- Documenting prognostic scoring
- Discussing cases in MDT